Medicine Quiz 1

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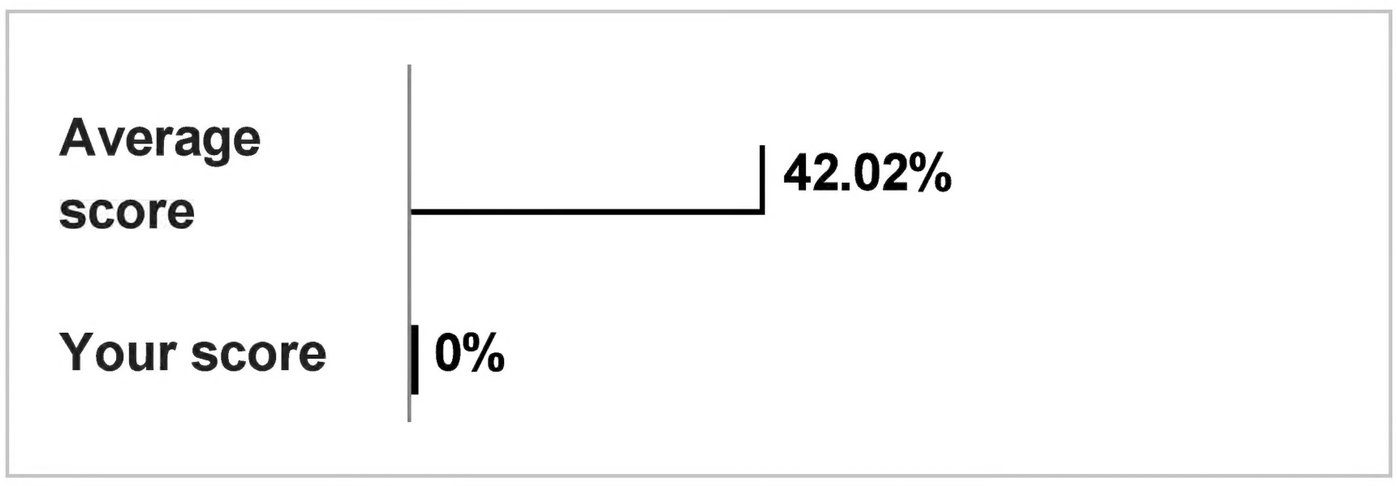

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1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16
17	18	19	20	21	22	23	24	25	26	27	28	29	30	31	32

Answered Review

1. Question **1 points**

An 85-year old woman who lives in a small rural Midwest town has always kept active despite her age, singing in her church choir, baby sitting her great-grandchildren, etc. However, for the past several months she has not felt well, not even able to muster sufficient energy to go to church on Sunday mornings, let alone go to choir practice. Also, she has problems sleeping, is extremely nervous with hand tremors, constantly anxious, sweats more than usual and can't stand having the heat on, has chronic diarrhea, and has the sensation that her heart races. When she mentions some of these observations to her local primary care provider, he basically shrugs them off, saying that no one of her age can expect to feel as they did when younger. He was worried however by the fact her systolic pressure rose from an average of 125 mm Hg to 185 mm Hg at the time of her visit; he prescribed hydrochlorothiazide and suggested she return in 3 months for a follow-up examination. However, in the meantime, she started to have trouble with her eyes: she had double vision and they appeared to her to be reddish and swollen; they also were itchy. Consequently her granddaughter took her to an optometrist who practiced in a nearby town. After her initial examination, the optometrist became alarmed to such a degree that she arranged a visit to the Mayo Clinic in Rochester Minnesota. After giving her a thorough physical, the physicians there started her on a therapy with the aim of alleviating the underlying cause of her symptoms, including the problem with her eyes. Which of the following choices most probably describes an aspect of the treatment used to alleviate the underlying cause of her symptoms?

1. Prescription of levothyroxine
2. Injection of interferon-β 1a
3. Watchful waiting
4. Prescription of zolpidem
5. Prescription of propylthiouracil ✔

INCORRECT ✖

The correct answer is 5.

This lady has Graves's disease and the optometrist recognized signs of Graves's' ophthalmopathy. Graves's disease is a form of hyperthyroidism caused by an autoimmune reaction of unknown etiology. The core of treatment is to reduce the production of thyroid hormone by the gland. This can be done by antithyroid drugs, radioactive iodine treatment, or surgery. Usually, the first line of treatment is by drugs either propylthiouracil or methimazole is used. The problem with this lady's eyes also needed to be treated with surgery to relieve the pressure behind the eyes caused by the infiltrating exophthalmos, which causes the swollen, bulging eyes very often associated with Graves's disease. It is thought that this exophthalmus is due to common antigens shared by the tissues behind the eye and the thyroid gland; as a result exophthalmus is an almost 100% specific symptom for Graves's disease. It is estimated that the incidence of Graves's disease is about 30–80 cases per 100,000 individuals, and it affects seven to eight females for every one male. Typically, the incidence peaks between the ages of 20–40 years. It is likely that this lady's primary health

care provider did not think Graves's disease because she was so far outside of the "typical" range. Had the disease not been discovered and treated it would have continued to ravage her body, possibly even leading to irreversible blindness and/or death.
(Choice 1) is incorrect. A prescription of levothyroxine would be used to treat a case of hypothyroidism, not hyperthyroidism. The most common cause of hypothyroidism is Hashimoto's thyroiditis; in adult women, the incidence is about 3.5 per 1,000 persons per year, and in men it is about 0.8 cases per 1,000 persons per year (but the incidence in both sexes is much higher in the Appalachian region). Like Graves's disease, Hashimoto's thyroiditis is an autoimmune disease, but it causes hypothyroidism rather than hyperthyroidism; however, short intervals of hyperthyroidism may intervene, particularly at the initiation of the disease. It is usually diagnosed by the presence of signs and symptoms of hypothyroidism (such as weight gain, sluggishness, sensitivity to cold, and depression), and confirmed by measuring the thyroid-stimulating hormone (TSH) level and presence of autoantibodies against various thyroid antigens. Chronic Hashimoto's can also cause goiter.
Interferons are a class of cytokines produced by many cells in response to double stranded RNA. They may aid the immune response by inhibiting viral replication within infected cells; they may also activate natural killer cells and macrophages and facilitate antigen presentation to lymphocytes.
(Choice 2) is incorrect. Although interferons β 1a and β 1b have anti-autoimmune activity that has made them useful in treating multiple sclerosis they have no known function in treating Graves's disease.
(Choice 3) is incorrect. Watchful waiting is defiantly not an acceptable treatment for Graves's disease; morbidity will only progress with time, perhaps even resulting in death.
(Choice 4) is incorrect. Zolpidem is used to treat insomnia. Although it might be used to treat the insomnia associated with hyperthyroidism, it will not alleviate the underlying cause of her symptoms.

2. Question **1 points**

A 67-year-old man presents with headache, vomiting, blurred vision, difficulty with speaking and swallowing, and loss of balance. He also complains of numbness in his face. His past history is significant for diabetes mellitus and hypertension. The patient presently is on metformin for the former and a combination of an angiotensin converting enzyme (ACE) inhibitor and diuretic for the latter. The acute problem occurred approximately 2 hours prior to being seen in the emergency room. His blood pressure is 150/100 mm Hg, his pulse rate is 80/min with a few irregular beats, and his temperature and respiratory rate are normal. The patient is conscious, the left pupil is smaller than the right, and the eyeball appears to be sunken into the orbit. He also has partial closure of the left upper eyelid. He has no pain sensations over the left side of the face to pinprick. However, touch is preserved. His gag reflex is depressed, and the palate is noticeably lowered on the left side, with the uvula being pulled to the right when he was asked to say, "aaah." There was a noticeable nystagmus to the left, and he has poor coordination of his left upper and lower extremities. His gait is ataxic. When tested with a pin, he cannot feel pain in the right upper and

lower extremities. However, the sensation of touch is preserved. A noncontrast computerized tomography (CT) of the brain was normal. Which of the following is the best means of immediate medical management in this patient?

1. Heparinization and observation
2. Right carotid endarterectomy
3. Left carotid endarterectomy
4. Thrombolytics ✔
5. Antiplatelet agents

INCORRECT ✖

The correct answer is 4.

This patient most likely has had an infarction involving the posterolateral portion of the medulla oblongata as a result of blockage of the posterior inferior cerebellar artery (PICA) on the left side. The condition is also known as Wallenberg syndrome, which might also be caused by occlusion of the vertebral artery. The computerized tomography scan shows no evidence of hemorrhage, and therefore the diagnosis of ischemic infarction is appropriate. Since his condition has been present for less than 3 hours, it is prudent to use a thrombolytic agent **(Choice 4)** as the initial medication. In the thrombolytic system, plasminogen is activated into plasmin by tissue plasminogen activator (tPA); plasmin then further decreases fibrin into degraded peptides. Plasminogen activator must be administered within 3 hours of the onset of the acute event to be efficacious. It is contraindicated if the blood pressure is sustained above l85/ll0. No other antithrombotic treatment should be instituted for 24 hours.
(Choice 1) followed by warfarin is appropriate in the management of a patient with a non-hemorrhagic stroke and would have been the choice had it been more than 3 hours after the onset of the acute event.
(Choices 2 & 3) Right or left carotid endarterectomy are incorrect, because the problem involves the vertebral circulation.
(Choice 5) Antiplatelet agents, such as aspirin and clopidogrel, are not used in the initial management of infarctions. They have a role to play in the prevention of strokes.

3. Question **1 points**

A 60-year-old man presents to the emergency room with a history of vomiting, increasing confusion, hearing voices, and blurred vision. The patient provided a history of congestive cardiac failure and reported he is being treated with an unnamed diuretic, enalapril, carvedilol, and digoxin. His blood pressure is 140/95 mm Hg; pulse, 88/min irregular; respirations, 22/min; and body temperature is normal. Physical examination confirmed altered mental status, an irregular cardiac

rhythm to auscultation, and basilar rales on the left, but there is no cyanosis or pallor. The electrocardiogram shows left axis deviation and paroxysmal atrial tachycardia with block. A chest x-ray film confirms left ventricular hypertrophy with blunting of the left costophrenic angle and diffuse shadowing at the base. Effective management of cardiac dysfunction would include which one of the following?

1. Raising the serum sodium level
2. Raising the serum magnesium level
3. Lowering the serum magnesium level
4. Raising the serum calcium level
5. Raising the serum potassium level ✔

INCORRECT ✖

The correct answer is 5.

This patient has symptoms of digitalis toxicity in addition to cardiac failure. Digitalis toxicity leads to vomiting, increasing confusion, hallucinations, photophobia, yellow vision, and cardiac arrhythmias that could be potentially dangerous if left untreated, because supraventricular tachycardia (SVT) and atrioventricular (AV) block could supervene, or even worse, ventricular tachycardia and fibrillation. The presence of SVT and AV block signifies cardiotoxicity. Digitalis toxicity may be precipitated by hypokalemia, and therefore, the serum potassium level should be increased, maintaining it between 4.0 and 5.0 mmol/L. There is a strong possibility the patient is on a loop diuretic (such as furosemide), which restricts sodium and chloride reabsorption in the proximal part of the ascending loop of Henle, with resultant excretion of sodium, water, chloride, and potassium. Digitalis administration should be discontinued temporarily. The serum digitalis level does not correlate with digitalis toxicity.

(Choice 1) will have no effect on the heart. Sodium has its primary effects on the brain and not the heart.

(Choice 3) would aggravate the problem further, as one of the causes of digitalis toxicity is hypomagnesemia.

(Choices 2 & 4) Raising the serum calcium level would be disastrous, as hypercalcemia promotes digitalis toxicity. Although hypomagnesemia could result from use of loop diuretics, this effect is less common than that of hypokalemia; therefore, raising the serum magnesium level is not indicated. Other causes of digitalis toxicity include hypothyroidism, myocardial ischemia, advancing age, renal insufficiency, volume depletion, and concomitant use of drugs that delay elimination of digitalis, such as amiodarone, verapamil, β-blockers, such as carvedilol (which has been prescribed to this patient) and quinidine, a cinchona alkaloid that is used to treat cardiac arrhythmias. Although with coadministration of angiotensin-converting enzyme (ACE) inhibitors, such as enalapril, digitalis toxicity can potentially induce hyperkalemia, in the case presented, the electrocardiogram did not show evidence of hyperkalemia (peaked T waves); if it did, one would consider lowering the serum potassium level.

4. Question

1 points

A 69-year-old man with a known history of coronary artery disease (CAD), atrial fibrillation, and hypertension presents with sudden onset of right facial weakness and numbness. He also complains of a roaring in the right ear. On examination, he has some difficulty with speech. He does not have a pronator drift, and his grip strength is normal. However, there is weakness of the face on the right side, including the orbicularis oculi. He is unable to appreciate taste on the anterior tongue on the right side. He has normal sensations on the face to touch and pinprick. Which of the following would best explain these findings?

1. Upper motor neuron lesion
2. Brainstem glioma
3. Left middle cerebral artery embolus
4. Hemorrhage within the left internal capsule
5. Lower motor neuron lesion ✔

INCORRECT ✖

The correct answer is 5.

This patient has a lower motor neuron lesion affecting cranial nerve VII—the facial nerve (CN VII) on the right side—and is also known as Bell's palsy. Facial palsy is the most common of the cranial neuropathies. Most cases are due to infection with herpes simplex virus and are not idiopathic, as was believed earlier. The facial nerve innervates the muscles of the face and the stapedius muscle in the ear and conveys taste fibers to the anterior two-thirds of the tongue, via the chorda tympani nerve. The roaring in the ear (hyperacusis) and lack of taste in the anterior portion of the tongue are due to involvement of the aforementioned innervation. The problem with speech is due to dysarthria, resulting from paralysis of the muscles around the mouth. The inability to close his eye can put his cornea, and indeed the eye, in danger. Hence, the eyelid should be taped shut or covered with an eye patch. The facial nerve also supplies the lacrimal gland; absence of tears could result in xerophthalmia (corneal drying) and attendant complications, hence artificial tears are necessary. Corticosteroids are helpful. The disorder usually resolves over time. Other causes of lower motor neuron facial paralysis include lesions in the brainstem, cerebellopontine angle, middle ear infection, multiple sclerosis, human immunodeficiency virus (HIV) infection, Lyme disease, parotid tumor tumors, diabetes mellitus, and trauma to the facial nerve.

(Choice 1) An upper motor neuron lesion of the facial nerve will spare the upper half of the face because of the bilateral innervation to that region. An upper motor neuron VII nerve paralysis is due to involvement of the contralateral motor neurons of the descending

corticobulbar pathway or of the pathway used en route to the seventh cranial nerve nucleus in the pons.
(Choice 2) is unlikely, given the sudden nature of the event, lack of symptoms such as headache, nausea and vomiting, and bulbar signs involving the lower cranial nerves. These patients could have ataxia as well.
(Choice 3) could very well occur given his history of atrial fibrillation. However, embolism involving the middle cerebral artery would be associated with headache, confusion, right hemiparesis or hemiplegia, and an upper motor neuron facial paralysis on the right as well.
(Choice 4) could occur, given the history of hypertension in this patient. However, the patient would have severe headache, obtundation, and a dense right hemiplegia, together with a right upper motor neuron facial nerve paralysis.

5. Question **1 points**

A 55-year-old woman presents to the emergency room with a history of severe crushing retrosternal chest pain while she was moving some furniture around the house. Electrocardiography shows elevated ST segments in leads II, III, and aVF, consistent with an inferior myocardial infarction. This diagnosis is further supported by cardiac enzymes that test positive. The patient is hemodynamically stable. She is moved to the intensive care unit. A few hours later, she developed tachyarrhythmia. Treatment with intravenous (IV) boluses of lidocaine controls the arrhythmia only transiently—the arrhythmia disappears within 1 minute only to reappear within 4 minutes of each bolus dose. Plasma levels of lidocaine measured soon after each injection remained within the therapeutic range (1–5 mg/L). Which of the following statements is most accurate?

1. Rapid metabolism of lidocaine is responsible for the short duration of its antiarrhythmic action after the bolus administration.
2. Laboratory determinations of blood levels of lidocaine after IV administration are frequently in error.
3. The elimination half-life of lidocaine is approximately 2 minutes.
4. Lidocaine rapidly redistributes from blood to other tissues. ✔
5. Cardiac cells rapidly develop tachyphylaxis to lidocaine.

INCORRECT ✖
The correct answer is 4.
Rapid (but temporary) control of arrhythmia follows achievement of a therapeutic blood level of lidocaine. The temporary nature occurs because lidocaine rapidly redistributes from blood to other tissues. In this situation, the reappearance of the arrhythmia reflects the rapid

distribution of lidocaine from the blood to highly perfused body tissues, resulting in a decrease in plasma concentration of the drug below therapeutic levels. A similar process, involving redistribution of thiopental from the brain to other highly perfused tissues, is responsible for termination of the anesthetic effects of the intravenous (IV) barbiturate.
(Choices 1 & 3) Although the liver metabolizes lidocaine extensively, the elimination half-life of the drug is 1-2 hours (not 2 minutes), which cannot account for such a rapid reappearance of the arrhythmia.
(Choice 2) While the blood levels following IV administration of lidocaine may be variable, this is due to redistribution processes and not to errors in laboratory determination of drug levels.
(Choice 5) There is no evidence that cardiac cells are capable of rapid (and reversible) changes in sensitivity to any antiarrhythmic drug. Conversely, the lidocaine level may be increased in patients taking cimetidine, and the dosage needs to be reduced in patients with congestive cardiac failure or liver disease.

6. Question

1 points

A 58-year old male computer programmer presented to his physician for a routine check-up after his wife nagged him into it. This was his first visit to a physician in at least a decade. After work, he would come home, eat dinner, watch television, and go to bed. He spent his weekends watching sports on television. While doing so, he often ate peanuts, buttered popcorn, or potato chips and downed it with a couple of beers. When asked if this had always been his lifestyle, he replies that prior to turning 40 he played sports and enjoyed dancing with his wife. His subsequent physical examination and laboratory work did not uncover any abnormalities with his heart, lungs, thyroid hormone levels, or kidney function. On the other hand, three different blood pressure readings taken by both the physician and his assistant obtain similar results, namely, an average reading of 178/92 mm Hg. He is 5 feet, 9 inches (1.75 m) tall and weighs 204 pounds (92.5 kg). A fasting blood sugar and lipid profile show a glucose level of 124 mg/dL, total cholesterol (TC) level of 234 mg/dL, and low-density lipoprotein (LDL) level of 158 mg/dL. The most appropriate first line of management in this patient would be which of the following?

1. Gastroplasty
2. Prescription of an anorexic drug
3. Prescription of gemfibrozil
4. Persuade him to take his wife out dancing at least twice a week. ✔
5. Prescription of a dietary supplement containing ephedra
6. Prescription of tolbutamide

INCORRECT ✖

The correct answer is 4.

This patient is in the early stages of metabolic syndrome and increases insulin resistance, which, when fully developed, is characterized by type 2 diabetes, dyslipidemia, hypertension, and obesity. Although only tested once, at this time, it is highly probable that he is in a pre-diabetic state, which is defined as having fasting plasma glucose levels between 100 and 126 mg/dL on two consecutive days. His fasting total cholesterol (TC) level is also in the borderline high range. (For TC, the normal value is defined as equal to or less than 200 mg/dL [5.2 mmol/L], borderline is between 200 and 240 mg/dL [5.2–6.2 mmol/L], and elevated as equal to, or greater than 240 mg/dL [6.2 mmol/L]. Whereas the new American Heart Association definitions for LDL levels are: optimal less than 100 mg/dL [2.5 mmol/L], near optimal 100–129 mg/dL [2.5–3.3 mmol/L], borderline high 130–159 mg/dL [3.4–4.0 mmol/L], high value 160–189 mg/dL [4.1–4.8 mmol/L], and very high 190 mg/dL [4.8 mmol/L] and above.) In addition, he is definitely hypertensive (systolic/diastolic values reported as mm Hg are defined as normal, <120/80; pre-hypertension, 120/80 to 139/89; stage 1 hypertension, 140/90 to 159/99; and stage 2 hypertension, ≥ 160/100). In summary, he is well on his way to becoming a full-blown diabetic, with a high blood glucose level as well as high LDL and total cholesterol levels, and also is liable soon to be a stage 2 hypertensive and a candidate for severe coronary artery disease (CAD). The primary driving force for these conditions is his obesity, created by his lifestyle. His body mass index (BMI) is 30.2 kg/m^2, calculated using the following formula:

BMI = weight in kg divided by height in meters squared, where 1 kg = 2.2046 lb, and 1 m = 39.37 inches.

A BMI between 25 and 29.9 kg/m^2 is defined as overweight, and 30 kg/m^2 or greater as obese. The first step in managing obesity in this patient is to get him off his couch and moving about. Taking his wife out dancing at least twice a week, while not a solution in itself would be a step in the right direction, and it might be easier to persuade this obviously recalcitrant man to do this, which may also please his wife, than to initially recommend the more stringent dietary and exercise regimes that will be required. In the interim, it also may be possible to persuade him to return to treat his hypertension and incidentally better deal with his lifestyle issues, including the possibility of depression.

(Choice 1) will cause him to lose weight. However, it requires surgery and has concomitant short and long-term risks. As a consequence, it should be reserved only for the morbidly obese.

(Choice 2) Although short-term weight loss is enhanced by the prescription of an anorexic drug, patients taking such agents usually end up even heavier than before they started treatment because of a rebound effect. Management in this case clearly entails a lifestyle change, which may require long-term treatment and a great deal of patience.

(Choice 3) One would prescribe gemfibrozil as a way to lower triglyceride levels only after they were acutely high and more conservative measures failed.

(Choice 5) A dietary supplement containing ephedra would not be a wise move since several studies have shown that it can cause serious reactions including hypertension, which certainly would not be recommended in this case. Ephedra (also known as ma huang) contains the chemical ephedrine, which was used as an asthma medication until the 1980s, when it was taken off the market because of its dangerous effects on the heart and blood

pressure. Tolbutamide is a first-generation sulfonylurea, an oral hypoglycemic agent. Not only are more effective agents available, but also tolbutamide, like all sulfonylureas, runs the risk of inducing hypoglycemia and promotes weight gain.
(Choice 6) The physician would not likely prescribe tolbutamide. Until recently, prediabetes would only be treated by lifestyle changes, diet, and exercise. However, it now is deemed acceptable to be more aggressive and prescribe an oral hypoglycemic agent. Metformin is often recommended because it neither causes hypoglycemia nor promotes weight gain.

7. Question **1 points**

A 44-year-old woman consulted her primary care physician because of a nagging feeling of fatigue, pain, and stiffness in both hands upon rising; the pain persisted for at least 1 hour after arising and was severe enough to make getting herself ready for the day's work difficult. The physician found that she had a low blood count, an elevated sedimentation rate, and an elevated C-reactive protein level, but x-rays of her hands were essentially normal. Feeling that she may have some sort of autoimmune condition, he referred her to a rheumatologist, who immediately had her haplotype determined and ran immunological tests for rheumatoid factor and an anti cyclic citrullinated peptide antibody (ACPA) test, as well as testing for antinuclear antibodies (ANAs). Her haplotype serotype was HLA-DR4, and she tested positive for ANA and ACPA and negative for rheumatoid factor. Most likely, this lady suffered from which one of the following conditions?

1. Systemic lupus erythematosus
2. Multiple sclerosis
3. Sjögren syndrome
4. Diabetes mellitus type 1
5. Rheumatoid arthritis ✔

INCORRECT ✖
The correct answer is 5.
The lady has the early symptoms of rheumatoid arthritis (RA). RA is an autoimmune inflammatory condition principally involving the synovial membranes of multiple joints, typically in a symmetrical bilateral fashion starting with the smaller joints on the hands. As the disease progresses, typically with periods of remission, it affects the feet and bigger joints, and causes malaise, weight loss, and fever. Although in this case the lady's symptoms and early laboratory studies are nonspecific, they are typical for some sort of inflammatory autoimmune condition. Moreover the HLA-DR4 haplotype serotype is strongly positively correlated with RA and a positive anticyclic citrullinated peptide (ACPA; also known as citrulline antibody, anticitrulline antibody, anti-CCP, and cyclic citrullinated peptide antibody)

test. This test measures a form of serum citrulline, and some investigators hypothesize that the conversion of arginine to citrulline plays a role in the autoimmune inflammatory process associated with RA. It has a specificity of about 95% for RA, although it does provide false-negative results about 33% of the time; in addition, it often is present in the earliest phases of the disease, even before clinical signs are clearly manifested. In contrast, the negative rheumatoid factor result does not rule out RA because it is frequently negative in early cases, even after the first year. ANA is a relatively nonspecific test for autoimmune and other inflammatory conditions; therefore, a positive test is compatible with RA but also other disease states. The prevalence of RA is about 1 case per 100 persons; females are affected three times more often than males, and the onset in most cases occurs between the ages of 25 and 50, peaking between the ages of 40 and 50 years. Recent treatments for RA have improved the lives of RA patents to a remarkable extent. Although the classic medications, corticosteroids and/or nonsteroidal anti-inflammatory drugs (including aspirin), are still employed to reduce pain and inflammation, so-called disease-modifying antirheumatic drugs (DMRDs) are now available and often prescribed early in the course of the disease to avoid disease progression to the point of causing irreversible damage. In addition, genetically engineered drugs called biologics have been developed that induce remissions in about two-third of cases. Unfortunately, these have to be administered by injection and are very expensive, thus they are often reserved for people in whom DMARDs are ineffective, or they are used to supplement DMARD treatments. Physical therapy is also an important adjunct used to help joint function.

All the incorrect choices also are autoimmune diseases.

(Choice 1) Systemic lupus erythematosus (SLE) is a chronic inflammatory systemic disease that can affect any organ system. ANAs are present in the serum of most patients with active SLE, and antibodies to native double-stranded DNA are essentially diagnostic for this disease. A genetic component is also likely, since HLA-DR2 and HLA-DR3 human leukocyte antigens are more common than in the general population. SLE is erratic in its manifestations in that it follows a relapsing and remitting course. The incidence is highest among women of childbearing age, and overall in adults, it is 10–15 times higher in women than in men. Cases of SLE vary from being relatively benign to being progressive and rapidly fatal. Prior to 1955, the 5-year survival was less than 5%, but now thanks to better medical care, the 10-year survival is about 90%. Nonetheless, 30% of SLE-related deaths occur before the age of 45 years.

(Choice 2) Multiple sclerosis (MS) is a disease in which the immune system attacks the myelin sheets in the central nervous system, killing oligodendrocytes and causing demyelination. This may produce numerous physical and mental symptoms, depending on the nerves affected, and it often progresses to physical and cognitive disability. Disease onset usually occurs in young adults, and it is more common in women. It has a prevalence ranging between 2 and 150 per 100,000 depending on the country or specific population; it is more frequent among individuals of northern European descent. Although the condition is not curable, recently developed medications help mitigate symptoms; the type of medication used varies with the variant form of the disease.

(Choice 3) Sjögren syndrome occurs when the immune system attacks and destroys moisture-producing organs, including the salivary and lacrimal glands and more rarely the lungs, kidneys, or gastrointestinal tract. It usually is characterized by dry eyes and mouth, and sometimes by enlargement of the parotid gland. More than 90% of individuals affected by

Sjögren syndrome are women, and 90% of the time it is rheumatoid factor positive; it also may be associated with rheumatoid arthritis or other rheumatic diseases. Serum levels of ANA are also often elevated. Some people may experience only mild symptoms of dry eyes and mouth and are able to treat the disease symptomatically, while others have a more severe disease and must cope with blurred vision or recurrent oral infections and difficulties with swallowing and eating. Systemic problems can also occur including fatigue, joint pain, and even autoimmune tubulointerstitial nephritis leading to proteinuria, urinary concentrating defect, and distal renal tubular acidosis.

(Choice 4) Unlike diabetes mellitus type 2, type 1 is a disease with an autoimmune component triggered by some still not clearly identified factor. Approximately 85% of patients have circulating islet cell antibodies, and the majority also has detectable anti-insulin antibodies before receiving insulin therapy. Most islet cell antibodies are directed against glutamic acid decarboxylase within pancreatic B cells. Some theorize that there is a virally induced etiology. The Coxsackie virus is commonly suggested, although the evidence is inconclusive. Since not everyone infected by this organism gets diabetes type 1, it also is suggested that, in addition, there is a genetic component; evidence for this is provided by the fact that up to 95% of patients with type 1 diabetes express human leukocyte antigen (HLA) DR3 or HLA-DR4; in fact HLA-DQs are considered specific markers of type 1 DM susceptibility. Others claim cow's milk contains a triggering agent, which however never has been identified; this idea is derived from studies claiming a higher incidence among babies given cow's rather than breast milk after birth.

8. Question **1 points**

A man married a young woman who lost her vision at the age of 28 years. They had three children, two girls and one boy, all of whom started to become blind after the age of 20. All three of these children married sighted persons and had children. One daughter had two boys and a girl, all of whom became blind. The other daughter had a boy and girl; both of them lost their vision as well. The son had three boys and a girl, all of whom were sighted and had excellent vision as long as they lived. In the third generation, all the boys who remained sighted and those who lost their vision early married women who remained sighted until they died, and all their offspring also remained sighted well into old age. The daughter of the second-generation male who retained her sight married a normally sighted man, and their offspring could see throughout their lives as well. The second generation women who became blind married men who retained their sight but their offspring of either sex lost their sight at an early age. Which of the following inheritance patterns is most likely to be present?

1. Autosomal dominant
2. Autosomal recessive
3. Mitochondrial ✔
4. X-linked dominant

5. X-linked recessive

INCORRECT ✖

The correct answer is 3.

In this pedigree, all affected females transmit a trait that causes early vision loss to offspring of both sexes. However, affected males and normal individuals of both sexes do not transmit loss of sight to their offspring. This non-Mendelian pattern of inheritance is characteristic of conditions caused by a mutation in a mitochondrial gene, due to the fact that only females transmit mitochondria to the ovum. This particular family has Leber's hereditary optic neuropathy. Individuals who inherit this condition suffer precipitous vision loss, usually in their 20s, due to optic nerve degeneration. In different families with Leber's hereditary optic neuropathy, 11 different missense mutations in three different mitochondrial genes encoding respiratory chain subunits have been described. Although the ultimate phenotype, loss of vision, remains consistent, the rate of vision loss and the age of onset vary considerably, even among members of the same family. Such variability in the phenotypic expression of conditions transmitted by mitochondrial mutations is characteristic of these conditions and is called heteroplasmy. It occurs because each cell carries multiple mitochondria, of which on average only half will be affected, but transmission of mitochondria with normal and mutated genes will occur in a random manner. Because mitochondrial-DNA (mtDNA)-encoded proteins are primarily associated with electron transport and adenosine triphosphate (ATP) synthesis, tissues that depend upon the electron transport system are affected most.

(Choice 1) In an autosomal dominant disorder, either an affected male or female may transmit the mutant gene to a child of either sex, or the mutant trait will be expressed.

(Choice 2) In an autosomal recessive disorder, both parents must transmit a mutant gene to the gamete for the trait to be expressed. An offspring of either sex who inherits one mutant gene from either parent will be a carrier.

(Choice 4) In an X-linked dominant disorder, affected males will carry only one X chromosome, which will bear the mutated gene; this X chromosome will be transmitted to his daughters, in whom the trait is expressed since it is dominant. However, characteristically, the disease is more severe in hemizygous affected males than in heterozygous affected females. Since his sons only will inherit his Y chromosomes, they will not be affected, nor will they be carriers. Affected females almost certainly will have one X chromosome with the normal gene and one with the mutated gene; thus, they transmit the mutant gene to 50% of their daughters and to 50% of their sons. Since the trait is dominant, the mother as well as offspring of both sexes who inherit the affected chromosome will express the trait.

(Choice 5) In an X-linked recessive disorder, males carrying an X chromosome with a mutated gene will express the trait since they only have one X chromosome; females carrying an X chromosome on the other hand will be carriers but will not expresses the trait since the recessive trait will not be expressed in the presence of an X chromosome carrying the normal gene. Sons of affected males will be neither carriers nor affected because they inherit a Y, not an X chromosome; however, 100% of the daughters will be carriers because they will receive the affected X chromosome from their father and a normal one from their mother. Fifty percent of the daughters of women who are carriers will become carriers themselves, since there is a 50% chance they will inherit the affected X chromosome.

Although there is also a 50% chance that her son will inherit an X chromosome bearing the mutated gene, those who do will express the disease because that will be their only X chromosome.

9. Question

1 points

A 57-year-old woman has had a rather remarkable history. She was born to a poor immigrant family and worked her way through college as a bartender in a small night club where patrons enjoyed a "good smoke" along with their drinks. After college, she went to law school, passed the bar, got a job in a good law firm, and became a partner within a decade. Moreover, she married and has two children. All her life she has been active and full of energy, but about 6 months ago she began to fatigue early; 2 or 3 months ago, she noticed a shortness of breath and a loss of appetite and of weight. Several weeks ago, she developed a dry cough, and last week, she spat up sputum with streaks of blood. As a nonsmoker, she though herself essentially immune to lung cancer; however, the blood in her sputum alarmed her and she consulted a pulmonologist. He took a history, gave her a general physical examination, and took chest x-rays followed by a computed tomography (CT) scan. He reported back to her that he had both bad and good news: "The bad news is that you have lung cancer; the good news is that the cancer is located in a defined area near the center of the lung but only in the left lobe and at the root of the bronchus; this means it seems not to have spread and is a relatively slow-growing variant; this provides hope it can be successfully treated." Which of the following lung cancer variants does this lady most likely have?

1. Adenocarcinoma
2. Squamous cell carcinoma ✔
3. Bronchoalveolar lung cancer
4. Large-cell carcinoma
5. Small (oat) cell cancer

INCORRECT ✖

The correct answer is 2.

Lung cancer is the 5th most common cancer in Australia for both sexes. However, it is by far the leading cause of cancer death among both men and women. It is estimated that in 2011 there were 10,511 new cases resulting in 8,137 deaths from lung cancer. More people will die from lung cancer than from colon, breast, and prostate cancers combined. Typically, lung cancer is divided into two major categories, small-cell and non-small cell. This is because small-cell cancers grow rapidly, metastasize readily, and are scattered throughout the lung, making them essentially impossible to treat by surgery or radiation. On the other hand, before they mature and spread, non-small cell cancers tend to remain concentrated in a

localized area, making surgical or radiation treatment a possibility. Among the non-small cell lung cancers are adenocarcinomas, squamous cell carcinomas, bronchoalveolar lung cancers, and large-cell carcinomas. Squamous cell carcinomas are generally located in a defined area near the center of the lung, either on a bronchial branch or on a nearby major lobe; this matches the description of the cancer the case above. These cells are situated near the terminus of the bronchi; consequently, they are particularly susceptible to airborne irritants and carcinogens and cancers in this area are typically caused by smoking tobacco products. However, carcinogens do not have to be smoked in order to be breathed in, as for instance is the case of second-hand smoke, which likely happened to this lady while she worked as a bartender. Typically, the progression from early injury is one that takes years to achieve; cells progress from normal, to hyperplasia, through metaplasia and dysplasia, finally into a true neoplastic state, a carcinoma. In this lady, the original exposure caused a mutation, and the progression may well have been promoted over the years by further exposure, likely during conferences, etc. Prior to the popularity of filter-tipped cigarettes, squamous cancers were the most common type of lung cancer, and they still constitute 25%–30% of all lung cancers. If caught in time, as it appears it may have been in the case described, squamous cell carcinomas are possibly treatable by surgery and/or radiation; however, they will grow and form bronchial cavities and thus become inoperable. It is the cancer type most often associated with hypercalcemia.

(Choice 1) usually occurs in a peripheral location in the lung and has become the most common form of non-small cell carcinoma, accounting for 35%–40% of all lung cancers. When contained, it tends to respond better than other lung cancers to surgical removal. This type of cancer is the most frequent form found in nonsmokers, but the relatively recent increase in its incidence is primarily due to smokers of low-tar filter-tipped cigarettes. Apparently, microscopic fragments from the filters imbedded with carcinogen from the cigarette are responsible for this cancer variant, which also is responsible for the rapid increase of this cancer among women and in individuals younger than 45.

(Choice 3) is a distinctive subtype of typical adenocarcinoma of the lung. A National Institute of Cancer survey found it differs from the typical adenocarcinoma in that it has a significantly higher 5-year survival rate among patients in all stages of the disease. Moreover, as compared to typical adenocarcinoma, bronchioalveolar carcinoma has a higher incidence of metastases within the lung but fewer to the brain. Bronchioloalveolar carcinoma also has been diagnosed with increasing frequency over the past few decades and now account for 3%–4% of cases of non-small cell lung cancer;. A characteristic finding in persons with advanced disease is voluminous watery sputum.

(Choice 4) cells are unusually large when seen under the microscope. In a chest x-ray, they usually appear in the central portion of the lung. They tend to be accompanied by extensive bleeding and tissue damage and to be undifferentiated, without the specific architecture found in other types of cancer cells. They grow quickly and metastasize at an earlier stage than other forms of non-small cell lung cancers. They also tend to induce paraneoplastic phenomena, including secretion of a hormone-like substance that causes gynecomastia in males.

(Choice 5) differ from the non-small cell carcinomas in that the are more aggressive, grow rapidly, metastasize to distant sites earlier, and are more frequently associated with distinct paraneoplastic syndromes. Except in rare situations, surgery plays no role in their treatment; however, they are very sensitive to chemotherapy or radiation.

10. Question **1 points**

A 56-year old woman presents at a family physician's office complaining of a chronic cough. She says she has had a cough for about the past 10 years, but it usually has been only during a brief period after waking up, especially on cold damp mornings. She confides she also is prone to catch chest colds in the winter but recovers without any problem. However, for the past 6 months or so, the cough has gotten worse. It lasts well into midday or later, and she hacks so badly that she can hardly work. Her bosses at the fan manufacturing plant where she has worked for the past 25 years have threatened to fire her unless she can get her all too frequent times of coughing under control; she adds "he also is pissed off because I miss so many days of work since I have so many colds." She continues, "My coughing sometimes is dry but more often produces a thick snotty-like spit which sometimes contains pus." When prodded, she admits she had been smoking since she was a teenager, and that for the last 10–20 years she probably smoked about two packs a day; she adds, "since cigarettes have became so expensive she has tried to cut back but can't because that long puff feels so good."

On physical examination, her blood pressure was 165/85 mm Hg. Her pulse is 100 and regular. Her weight is 185 pounds (83.9 kg), height 5 feet, 6 inches (2.2 m), and her body mass index (BMI) is 29.9. She wheezes while talking. On auscultation, adventitious breath sounds are heard in all lobes. X-ray demonstrates significant bronchial wall thickening and increased markings at the base of both lungs. Which is the most likely diagnosis in this patient?

1. Emphysema
2. Allergic bronchitis
3. Asthma
4. Chronic bronchitis ✔
5. Smoker's cough

INCORRECT ✖

The correct answer is 4.

Chronic bronchitis is a recurrent problem defined by coughing up an abnormal quantity of viscous mucus nearly every day for at least 3 months of the year for two or more consecutive years, clearly this lady meets this definition. In chronic bronchitis, the mucociliary response that normally clears bacteria and mucus from the bronchia is inhibited by damage to the endothelium caused by pollutants, primarily those caused by cigarette smoke. The inability to clear bacteria efficiently makes these persons vulnerable to catching colds, as described for the case described. The inflamed endothelium, combined with loss of supporting alveolar attachments, allows airway walls to deform and narrow the airway lumen;

this, plus the excess phlegm, obstructs the bronchia, causing the obstructive component of chronic bronchitis. The body responds by decreasing ventilation and increasing cardiac output, resulting in a rapid circulation of hypo-oxygenated blood. This produces hypoxia and eventually polycythemia, hypercapnia, and respiratory acidosis; these effects in turn cause pulmonary artery vasoconstriction, cor pulmonale, and signs of right heart failure. The difficulty these patients have in exhaling caused by the blockage of the bronchia tends to make these patients purse their lips, and the hypoxia makes the lips take on a blue-ish tinge. Consequently, persons with long-term chronic bronchitis are called “blue bloaters.”

(Choice 1) is defined as an abnormal, permanent enlargement of the air spaces distal to the terminal bronchioles accompanied by destruction of their walls without obvious fibrosis. It is believed that noxious irritants found in cigarette smoke (and less often to other pollutants) activate polymorphonuclear leukocytes and macrophages, which release human leukocyte elastase and other proteases; these eventually overwhelm the anti-proteases of the lung, resulting in tissue destruction. As the alveolar septae and the pulmonary capillary bed are eroded, the ability to oxygenate circulating blood is diminished; the body compensates for these events by reducing cardiac output and hyperventilating. This results in a limited blood flow through a fairly well oxygenated lung. Because of low cardiac output and the collapse of the alveoli, the rest of the body suffers from tissue hypoxia and generalized cachexia. Eventually, these patients develop muscle wasting and weight loss and can be recognized as “pink puffers.” Both chronic bronchitis and emphysema lead to chronic obstructive pulmonary disease (COPD), an irreversible chronic disease state caused by obstructive air flow. Although most cases display characteristics of both chronic bronchitis and emphysema, it has been estimated that chronic bronchitis predominates in 85%–88% of the cases.

(Choice 2) refers to short-term inflammations of the tracheobronchial tree caused by infections, allergens, or irritants. Although found in all age groups, bronchitis is diagnosed most frequently in children younger than 5 years.

(Choice 3) In asthma, the airways narrow as they become inflamed, constrict, and produce excess mucus in response to a triggering stimulant such as an allergen, an irritant, temperature changes, exertion, or emotional stress. These responses of the airways cause symptoms such as wheezing, shortness of breath, chest tightness, and coughing. Between episodes, patients usually feel well but they may develop shortness of breath more rapidly while exercising and retain it longer after exercise than the non-asthmatic individual. Symptoms of asthma range from mild to life-threatening but most often can be controlled by use of bronchodilators, oral drugs, and/or environmental changes. Asthma can be mistakenly diagnosed as acute bronchitis if the patient has no prior history of asthma. In one study, one-third of patients who had been determined to have recurrent bouts of acute bronchitis were eventually identified as having asthma.

(Choice 5) typically is an early morning cough demonstrated by most smokers. Nonsmokers clear harmful substances from their lungs via the action of cilia, which beat outward, thereby sweeping the bronchi clean. Smokers, however, both accumulate more toxic materials in their lungs than do nonsmokers and also cigarette smoke poisons the action of the cilia. Consequently, during the day, these toxic, irritating substances accumulate in smoker’s lungs. However, during the night, during which the smoker usually refrains from smoking,

some cilia recover and begin working again and after waking up, the smoker coughs, trying to clear away the poisons that built up the previous day. Unfortunately, prolonged smoking leads to the permanent destruction of the cilia and chronic bronchitis.

11. Question

1 points

A 22-year-old woman consulted a physician because she had been feeling poorly. She informed the physician that she is extremely tired despite going to bed early and sleeping well. This first became a problem almost a year ago, and more recently she also noted she became extremely weak after exerting herself physically or even after eating a carbohydrate-rich meal. Additionally, she was thirsty all the time, even though she also craved extra salt; understandably, she also needed to urinate excessively, even at night, thus interrupting her sleep. In response to the doctor's questions, she also revealed that these spells of weakness involved all four limbs and were worse in and around her upper arms and shoulders, as well as in her hip muscles. This made walking almost impossible, while her weak arms caused her to drop and break things. She added that she also has been getting severe cramps and painful spasms in these muscles. In addition, she thought that she has more joint stiffness, some degree of numbness in her hands and feet, and sometimes she felt dizzy upon arising. She had no history of nausea, vomiting, or other gastrointestinal problems. She added that she does not smoke, only drinks alcohol at a few social occasions, and is not taking any prescription or over-the-counter drugs; no person in her family that she was aware of had similar problems; however, her parents are first cousins. A "weakness attack" was induced by making her lift weights while marching in place and then having her rest. During the ensuing attack, the muscles in all four limbs became flaccid and there were associated depressed tendon jerks. The muscles of her eyes, face, tongue, pharynx, larynx, diaphragm, and sphincters were not involved. Her blood pressure remained within the high normal limit. Laboratory studies showed her serum glucose was 132 mg/dL, sodium, 140 mmol/L (normal 136-145), serum calcium 2.5 mmol/L (normal 2.2-2.6), serum potassium ion value was 1.6 mmol/L (normal 3.5-5.0), serum magnesium ion level was 0.5 mmol/L (normal 0.8-1.2), and serum chloride ion value was 94 mmol/L (normal 98-106 mmol/L); the abnormal levels persisted even between the attacks. She also had metabolic alkalosis with a bicarbonate ion level of 33 mmol/L (normal 22-30 mmol/L) and a blood pH of 7.48 (normal 7.35-7.45). A 24-hour urine analysis revealed alkaline urine with higher than normal amounts of potassium ion, 285 mmol/24h (normal 25-125 mmol/24h); chloride ion, 625 mmol/24h (normal 110-250 mmol/24h); and magnesium ion, 50 mmol/24h (normal 2-5 mmol/24h). On the other hand, the urinary calcium ion was lower than normal (1.5 mmol/24h, normal 2.5-7.5/24 hr). Thyroid function tests (T3, T4, and TSH) were normal. Which of the following does this lady most likely have?

1. Thiazide diuretic toxicity
2. Bartter syndrome
3. Dent syndrome
4. Andersen-Tawil syndrome

5. Gitelman syndrome ✔

INCORRECT ✖

The correct answer is 5.

Gitelman syndrome is a disease of the kidney's distal convoluted tubule that results in the loss of magnesium, sodium, potassium, and chloride ions into the urine. Most cases are either isolated or inherited in an autosomal recessive fashion, although there is at least one reported family in which the disease was reported to be inherited in an autosomal dominant manner. The most common cause of Gitelman syndrome is a mutation in SLC12 A3 gene, although a few cases have been attributed to aberrations in CLCKKB. Several different mutations have been described in SLC12 A3 gene; this may account for differences in symptoms reported in various families. Most cases are relatively benign but some are more severe. The product of SLC12 A3 gene functions in the distal convoluted tubule as part of the thiazide-sensitive NaCl co-transporter, and homozygous loss of function mutations in this protein induces a hypocalciuria, but there is excess urinary loss of sodium, potassium, and magnesium ions. Hydrogen ion is retained, causing an alkaline urine. These changes are reflected by low circulating levels of potassium and magnesium ions, which are responsible for the symptoms of fatigue, muscle weakness, spasms, and pain. As a rule, the disease is not recognized until adolescence and often not until middle or even old age. A high-carbohydrate meal precipitates an attack because increased levels of glucose increases insulin levels, and the resultant increase in glucose utilization lowers circulating potassium ion levels. Treatment usually consists of replacement of potassium and magnesium ion. Some investigators also recommend an anti-prostaglandin such as oral indomethacin, which was found to assist in returning potassium ion levels to normal in at least one case.

(Choice 1) In so much as thiazide diuretics act on the same NaCl co-transporter as affected in Gitelman syndrome, it is not surprising that thiazide diuretic toxicity produces effects similar to Gitelman syndrome. However, in the case presented here, the patient states she does not take medication, consequently **(Choice 1)** is not correct.

(Choice 2) refers to a group of autosomal recessive conditions, all of which, like Gitelman syndrome, cause the loss of excess potassium and magnesium ions into the urine. At one time, Gitelman syndrome was thought of as a mild form of Bartter. However, they clearly are distinct entities. Bartter syndrome usually makes its presence know in neonates or early antenatals and essentially always before the age of 6 years; in contrast, Gitelman strikes adults. Urinary excretion of magnesium is high in Gitelman syndrome and within the reference range in Bartter syndrome; conversely urinary calcium ion levels are high in Bartter syndrome and low or within the reference range in Gitelman syndrome. The final clinching difference is that different genes are involved. Recent genetic analyses have shown that there are five Bartter syndrome variants, as summarized in the following table.

Bartter Type	Gene	Affected Site
Type I	SLC12A1	The Na-K-2Cl transporter in the distal convoluted tubule

Type II	ROMK1	Apical potassium channel
Type III	CLCNKB	Basal chloride channel in the thick ascending limb of Henle
Type IV	BSND	In barttina, a subunit protein of the basal chloride channel in the thick ascending limb of Henle
Type V	CLCNKA	In barttin, a subunit protein of the basal chloride channel in the thick ascending limb of Henle

(Choice 3) is a rare X-linked recessive disorder sometimes called idiopathic hypercalcuria. It is caused by mutations in the renal chloride channel CLCN5, and it causes low-molecular-weight proteinuria, hypercalciuria, aminoaciduria, and hypophosphatemia. Affected males will usually show symptoms in early adulthood, including tubular proteinuria, hypercalcuria, calcium nephrolithiasis, nephrocalcinosis and hypophosphatemic rickets; a significant percentage will have end-stage renal failure before the age of 50. Because of these properties, it is one of the several diseases that cause Fanconi syndrome.
(Choice 4) is an autosomal dominant disease characterized by a triad of symptoms:

1. Episodic flaccid muscle weakness (i.e., periodic paralysis typified by weakness that occurs spontaneously following prolonged rest or after rest following exertion)
2. Ventricular arrhythmias with a prolonged QT interval.
3. Physical anomalies that may include low-set ears, eyes set far apart, small mandible, fifth-digit clinodactyly, syndactyly, short stature, scoliosis, and mild learning disability.

Most commonly, serum potassium levels are reduced during periods of weakness. The disease is caused by mutations in KCNJ2, which codes for the inward rectifier potassium channel 2, known as Kir 2.1, which is involved in setting and stabilizing resting membrane potentials and is primarily expressed in skeletal muscle, heart, and brain.
Phosphatidylinositol 4, 5 bisphosphate (PIP2) is an important regulator of Kir2.1 and many KCNJ2 mutations alter PIP2 binding.

12. Question **1 points**

A 77-year-old man fractured his femur and was undergoing surgery to insert a rod to facilitate healing. After the fracture, the distal portion of the femur was pulled up toward the hip by the action of his muscles; this caused a lot of bleeding, fortunately, the femoral artery was not punctured. Now, during the surgery itself, more bleeding occurred, and it was decided to transfuse him with

two units of blood. About 90 minutes after the start of the transfusion, it was noted that the patient developed chills; a quick check of his temperature showed it rose by 1.5°C from a presurgical value of 37.8°C. Which of the following reactions is most likely occurring?

1. A hemolytic transfusion reaction
2. A nonhemolytic febrile reaction ✔
3. A delayed hemolytic transfusion reaction
4. An allergic reaction
5. A reaction due to volume overload
6. An acute lung injury
7. A reaction caused by increased oxygen affinity

INCORRECT ✖

The correct answer is 2.

A nonhemolytic febrile reaction happens during 3%–4% of transfusions, making it the most common type of transfusion reaction and thus the most likely reaction to have occurred in the case described. It is thought that the most likely cause of this reaction is the presence of antibodies against white blood cell human leucocyte antigen (HLA) in the donor blood. Another possibility is the release of cytokines from white cells during storage; however, this is most likely to happen in stored platelet preparations. Nonhemolytic febrile reactions occur most frequently in patients who have had blood transfusions before or in multiparous women. Such febrile reactions are successfully treated with acetaminophen, and some hospitals routinely give patients acetaminophen prophylactically prior to surgery and/or use pre-storage leukocyte-reduced blood components, at least for high-risk patients. Once fever is noted, the transfusion must be terminated and the blood rechecked to make sure that it truly is the proper type for the patient and the blood pack in question is destroyed. This is an expensive interruption, making it worthwhile to take such measures to avoid it when practical.

(Choice 1) is the most dangerous type of reaction and approximately 20 patients die each year because of it; it occurs in about 1 in 40,000 transfusions. ABO incompatibility is most common, although antibodies against Rh or other blood group antigens do occur. Most common causes are: mislabeling of the recipient's pre-transfer sample, giving the wrong blood to a patient with the same name, or not checking the patient's blood bank identification tag with the identification number on the unit; a laboratory error in identifying the proper blood type very rarely occurs. The severity of the reaction depends upon the degree of incompatibility, the transfusion rate, the amount of blood product given, and the health of the recipient. Most often, the onset of symptoms is within an hour of initiation of the transfusion, but it can be delayed. If the patient is under general anesthesia, the only symptoms of the reaction may be hypotension, profuse bleeding from incision sites and mucous membranes caused by disseminated intervascular coagulation, and/or a dark urine caused by

hemoglobinuria. If a hemolytic transfusion reaction is suspected, the transfusion must be stopped, the sample and patient identification rechecked, and supportive treatment begun. The initial goal of treatment is to maintain blood pressure and renal blood flow with IV isotonic saline and furosemide.

(Choice 3) may occur if the patient has been exposed to an antibody level so low that it gave a negative pre-transfusion test. In this case, the reaction may occur as late as 1–4 weeks after the transfusion. Usually, the reaction is not very severe, often limited to the transfused erythrocytes causing a drop in the hematocrit. If a more severe reaction does occur, it should be treated as if it were a hemolytic transfusion reaction.

(Choice 4) An allergic reaction to an allergen in the donor blood is not uncommon. Usually, the reaction is mild with urticaria, edema, and maybe dizziness during or immediately after the transfusion. An anaphylaxis reaction is rare, most often occurring in IgA deficient recipients. In the case of an anaphylactic reaction, transfusion should be stopped immediately and the reaction treated. Patients with IgA deficiency should be transfused with blood from an IgA-deficient donor.

(Choice 5) might be induced in susceptible patients by the high osmotic load of blood products in the transfusion, which draws water into the intravascular space. To reduce this possibility, red blood cells should be infused slowly. Susceptible patients include those with renal insufficiency or cardiac problems. The patient should be observed carefully, and if signs of problems arise, the transfusion should be stopped and the patient treated; typical treatment is with a diuretic.

(Choice 6) may be caused by anti-HLA or anti-granulocyte antibodies in the donor plasma that agglutinate and degranulate recipient granulocytes in the recipient's lung. Such a reaction occurs in 1 case per 5,000 to 10,000 transfusions. Although many cases are mild, the reaction may cause severe acute respiratory symptoms, even death, making this the second most common cause of transfusion-related death following ABO incompatibility. In nonfatal cases, supportive therapy leads to complete recovery, but even mild incidents should be reported. Blood stored for more than 7 days has a marked decrease in the red cell 2, 3-diphosphogylycerate (DPG) level, and after 10 days, the DPG level is essentially nil.

(Choice 7) This absence enhances the affinity of hemoglobin for oxygen, slowing the release of oxygen to the tissues of a recipient. Such an effect can be significant in infants, sickle cell patients, and patients suffering a stroke or acute chest symptoms.

13. Question **1 points**

A 65-year-old man sees his primary care physician because of pain in his chest that he thinks might indicate heart trouble. He describes the pain as a burning sensation in his chest that radiates into the neck and his left arm. He also reports that he regurgitates a sour-tasting liquid that leaves a feeling of "pins and needles" in his throat and mouth. These symptoms are not aggravated by exercise but are by drinking coffee or chocolate, or by eating fatty foods. Results of a physical examination that included his heart sounds and an electrocardiogram (ECG) taken as part of an

exercise tolerance test are unremarkable. An upper gastrointestinal barium x-ray study shows the presence of a hiatal hernia. Which of the following is the primary mechanism for this patient's complaints?

1. Decreased acid production in the stomach
2. Increased gastric emptying
3. Decreased esophageal peristalsis
4. Inappropriate relaxation of the lower esophageal sphincter (LES) ✔
5. Absence of ganglion cells in the myenteric plexus

INCORRECT ✖

The correct answer is 4.

Although these symptoms clearly indicate that the patient has gastroesophageal reflux disease (GERD), it is always a wise move to rule out the possibility of cardioarterial disease (CAD), as was done by the exercise tolerance test. GERD is a chronic, recurrent disorder characterized by heartburn, belching, and epigastric pain. The primary mechanism is inappropriate relaxation of the LES. Agents that decrease LES pressure, such as chocolate, coffee, ethanol, fat, peppermint, anticholinergics, theophylline, diazepam, barbiturates, and calcium channel blockers, exacerbate GERD. About 1 in 10 nonpregnant adults complain about heartburn at least once a week, and prevalence of GERD increases with age, is found more commonly in older men than in older women, and is very common during pregnancy; approximately 1 in 4 pregnant women experience heartburn from acid reflux daily during pregnancy. This very high frequency is attributed to impaired LES competence coupled to increased abdominal pressure.

Secondary factors that predispose to GERD include hiatal hernia; acid concentration of the refluxate; delayed acid clearance, which is associated with low-amplitude peristalsis and recumbency; and decreased gastric emptying, which aggravates reflux. The acid concentration of the refluxate is the most important factor in determining progression to reflux esophagitis. Complications associated with GERD are reflux esophagitis, stricture formation, and Barrett's esophagus. Barrett's esophagus is a premalignant condition characterized by glandular metaplasia of the distal esophagus. The risk of developing adenocarcinoma is 2%-10% in the United States, and distal adenocarcinoma of the esophagus has replaced squamous cell carcinoma of the esophagus as the most common esophageal cancer.

GERD can also precipitate an asthmatic attack by reflux vagal stimulation or aspiration. Approximately 80% of adults with bronchial asthma have GERD. This percentage appears to be independent of bronchodilator therapy. Chronic aspiration is also linked to pulmonary fibrosis and bronchiectasis. Ambulatory monitoring of esophageal pH for 24 hours, the gold standard, can detect GERD and provide information relative to the potential for chronic aspiration of acid reflux. The treatment of GERD involves reducing gastroesophageal reflux, neutralizing the acid reflux, enhancing esophageal clearance, and protecting the esophageal mucosa from ulceration. Diet modification, weight loss, postural therapy (elevation of the

head of the bed), restriction of alcohol, and cessation of smoking, along with pharmacologic treatment, are used in a stepwise approach reflecting patient response. A recommended progression of drugs includes antacids and alginic acid → histamine-2 (H_2) receptor antagonists at conventional doses → H_2 receptor antagonists at high doses, omeprazole, or prokinetic agents (metoclopramide) → antireflux surgery.
(Choice 1) is noted in pernicious anemia (destruction of the parietal cells) and Helicobacter pylori-induced atrophic gastritis involving the antrum and pylorus.
(Choice 3) is noted in both achalasia and systemic sclerosis.
(Choice 5) is the key finding in achalasia.

14. Question **1 points**

A 42-year-old woman complains of aching in the shoulder muscles, difficulty with climbing stairs, and difficulty with swallowing solids and liquids from her upper esophagus. Physical examination reveals atrophy of her shoulder and pelvic muscles. There is erythema over the bridge of her nose, with violaceous discoloration and edema of the upper eyelids. Erythematous papules are noted over the proximal interphalangeal joints, metacarpophalangeal joints, elbows, and knees. An electromyogram (EMG) reveals low-amplitude polyphasic potentials, and laboratory testing reveals an increased creatine phosphokinase (CPK). Which of the following is the most likely diagnosis?

1. Polymyalgia rheumatica
2. Rheumatoid arthritis
3. Polymyositis
4. Dermatomyositis ✔
5. Systemic lupus erythematosus (SLE)

INCORRECT ✖
The correct answer is 4.
The patient has dermatomyositis (DM), which has some overlapping features with polymyositis (PM). Both disorders are a female-dominant type of myositis, with DM involving skin and muscle in adults and children, and PM involving only muscle in adults **(Choice 3)**. Both disorders have muscle pain and atrophy, with the shoulders and pelvic muscles most commonly involved. The pelvic muscle weakness makes it difficult for patients to climb stairs. Dermatologic lesions associated with DM (not PM) include erythema over the bridge of the nose, with violaceous discoloration and edema of the upper eyelids; additionally, erythematous papular lesions may be noted over the dorsum of the proximal interphalangeal joints, knuckles, elbows, and knees. Dysphagia for solids and liquids is present, because of weakness of the striated muscle in the upper esophagus. Laboratory findings include a

positive serum antinuclear antibody test result in 30% of cases. Anti Jo-1 and anti PM-1 antibodies are present in a small percentage of cases. The serum creatine kinase is increased, and muscle biopsies show a lymphocytic inflammatory reaction with destruction of muscle fibers. The treatment for DM and PM is chronic prednisone therapy. If patients fail to respond to steroid therapy, they may be placed on immunosuppressive agents such as azathioprine, cyclophosphamide, or methotrexate.
(Choice 1) may be associated with proximal muscle aching; however, serum creatine and the electromyogram (EMG) are normal.
(Choice 2) Likewise, rheumatoid arthritis does not feature increased in serum creatine kinase or an abnormal EMG.
(Choice 5) could easily explain the “butterfly rash” but would not explain the myopathic features expressed in the EMG and the increased muscle enzymes.

15. Question **1 points**

A well-dressed but somewhat dishevel male presents at a walk-in clinic complaining of difficulty with breathing and a pain in his chest with a chronic cough. Upon providing a history, he says that he is a
31-year-old Harvard MBA graduate and admits that, although he is the CEO of a small business, he has been burning the candle at both ends for at least the past 5 years and has not taken care of his health. The attending physician notices his foul-smelling breath before anything else. The patient also indicates he has not been feeling well for a couple of weeks and is bothered by profound sweating, an inability to sleep, and has lost 5 pounds during the past month. Upon further questioning, he admits he has a drinking problem and more than occasionally drinks until he passes out. Upon examination, the doctor notices his teeth are in sad shape, and he has evidence of gingivitis. Additionally, his temperature was 37.5°C (99.5°F) and he has decreased breath sounds and crackles over the area where the pain was most intense; there is also egophony, and dullness to percussion in the presence of effusion. A chest x-ray was performed; it showed consolidation with a single cavity containing an air-fluid level in the posterior segment of the upper right lobes. Among the follow- ing choices, which represents the best next step in the treatment of this man?

1. A prescription for oral metronidazole
2. A prescription for oral clindamycin ✔
3. A prescription for oral vancomycin
4. A prescription for oral penicillin G
5. A prescription for oral isoniazid

INCORRECT ✖

The correct answer is 2.

The chest x-ray showed a fluid-filled cavity characteristic of an abscess. Characteristically, this is due to a necrotizing lung infection that produced a pus-filled cavitary lesion. Such abscesses are generally caused by aspiration of oropharyngeal material by patients who are unconscious. The risk is increased in patients with gingivitis or poor oral hygiene. This patient has poor oral hygiene and confessed to bouts of unconsciousness under the influence of alcohol. The foul smell of his breathe suggests anaerobic bacteria are at least partially responsible for the infection. In the absence of additional data, the best course of treatment is a broad-spectrum antibiotic such as clindamycin. Had he been sicker and if available in a walk-in clinic, he would most likely been given clindamycin IV. In any case, he should be advised to either see his own physician, or to return within a week; unfortunately, walk-in clinics are not designed to provide continuous, coordinated, or comprehensive care.

(Choice 1) has activity against anaerobes and protozoa, but to be used in this situation it must be combined with an antibiotic such as penicillin to make sure the growth of aerobes is also inhibited.

(Choice 3) A prescription for oral vancomycin would be used if infection by a methicillin-resistant Staphylococcus aureus (MSRA) organism were suspected; however, that is not the case, and it is best to reserve use of vancomycin for treatment of resistant bacteria, to minimize the development of vancomycin resistance.

(Choice 4) A prescription for oral penicillin G would not effective because it would be degraded by the stomach's acidity; consequently, it always is administered parentally. Moreover, it would not be effective against gram-negative bacteria.

(Choice 5) A prescription for oral isoniazid would be used to treat tuberculosis, which would be a very unlikely possibility in this patient.

16. Question

1 points

A 30-year-old woman, who is a nonsmoker, presents with bilateral puffiness and swelling of the fingers and joint pains. Cold exposure and stress cause episodes of blanching of the fingers. After turning white, these digits take on a blue-ish tint and then a red one. She is not on any medications, and her liver aminotransferase enzyme activities have always been stable and within the normal range. Her thyroid function tests are also normal. The mechanism for this patient's disease is most likely the result of which of the following?

1. Vasospasm and thickening of the digital arteries ✔
2. Hyperviscosity due to cryoglobulins
3. An immune complex vasculitis
4. Thrombosis of the digital vessels

5. An embolism to the digital vessels

INCORRECT ✖

The correct answer is 1.

The patient has Raynaud's phenomenon, which most commonly affects women aged between 20 and 50 years. The phenomenon is marked by the changing color of the digits, as described for this patient. The blanching is due to a diminishing blood supply; the blue tint followed by a reddish color is caused by prolonged hypo-oxygenation, followed by flushing when the blood vessels open again. The phenomenon is commonly seen in association with various rheumatoid conditions, including scleroderma, rheumatoid arthritis, and systemic lupus erythematosus. In these conditions, the symptoms of Raynaud's phenomenon may precede the other symptoms of the underlying disease by years or even decades. This most notably occurs in sclerosis; it has been reported to be the presenting sign in 85% of the cases of systemic sclerosis and in 95% of the cases of localized scleroderma (aka, the CREST syndrome). Similar symptoms can be induced by other factors including hypothyroidism, carcinoid, frostbite, chronic use of vibrating tools, or a side effect of several medications, or the underlying factor(s) may be idiopathic. In idiopathic cases or those not involving generalized physical aberrations, in particular a rheumatoid disease, the multi coloration of the digits is often referred to as Raynaud disease rather than Raynaud's phenomenon. This question asks which choice most likely describes the underlying factor causing Raynaud's phenomenon in the patient described, and the data presented essentially eliminate most nonrheumatic causes of the phenomenon; in most, if not all, of the underlying rheumatoid conditions, the responsible factors playing critical roles responsible for initiating these changes are vasospasm and thickened digital vessels.

(Choice 3) Although these rheumatoid conditions are autoimmune diseases, immune complexes are not present in the vessels.

(Choices 2 & 4) Two other systemic causes of Raynaud's phenomenon are thromboangiitis obliterans (Buerger disease), which is an inflammatory vasculitis producing thrombosis of the digital vessels in male smokers (patient is female and a nonsmoker); and cryoglobulinemia, with proteins that precipitate in cold temperatures. The latter condition is commonly associated with chronic hepatitis C (not present in this patient, as demonstrated by her normal, stable liver transaminase levels). In addition to affecting the digits, in cold weather this condition also produces cyanosis of the nose and ears that reverses to normal when returning to a warm environment.

(Choice 5) Embolism to digital vessels in the hand is uncommon, and when it does occur, it is most commonly due to infective endocarditis or atrial fibrillation, neither of which is present in this patient.

17. Question **1 points**

A 33-year-old man presents as a new patient to a primary care physician. He tells the physician that he has had almost constant abdominal pain for at least the past month, and he lost his appetite and about 15 pounds during the past month; he also complains of being fatigued almost all the time. In addition, he has had frequent bouts of bloody diarrhea and the diarrhea and the concomitant need to wipe his bottom left his rectum sore. He adds that he had a similar sequence of events about 5 years previously, and at that time, his physician told him he had a nervous stomach. He adds that his younger brother seems to be having similar problems. The physician notes that he looks pale, and upon examination, she finds he has diffuse abdominal tenderness with the right lower quadrant being the most sensitive area. His temperature is 99.5°F (37.5°C),blood pressure 98/78 mm Hg, pulse 90 and regular; a complete blood count reveals that his serum hemoglobin value is 9.8 g/dL and a peripheral blood smear reveals a decreased mean corpuscular hemoglobin count (MCHC) and a decreased mean corpuscular volume (MCV). Among the following choices, which most likely represents the disease that this man suffers from?

1. Crohn's disease ✔
2. Ulcerative colitis
3. Irritable bowel syndrome
4. Lactase deficiency
5. Nervous stomach

INCORRECT ✖

The correct answer is 1.

Crohn's disease is an inflammatory autoimmune condition with a significant genetic predilection; however, the course of the disease varies greatly from patient to patient. In many cases, attacks go into remission only to recur again years later; in other cases, the disease is consistent and persistent. The inflammation causes bowel cells to secrete large volumes of water with more salt than can be resorbed; consequently, it causes diarrhea. In mild cases, this may only be annoying but in severe cases patients may have dozens of bowel movements per day, interfering with daily activities and sleep. The inflammation also may cause thickening and scarring of the mucosa, which in turn causes cramping and pain. Again, the pain can vary from a minor annoyance to almost being intolerable. In addition, the inflammation can lead to ulceration and bleeding, which can induce a microcytic anemia, as in the case described, or less commonly it may interfere with vitamin B12 absorption in the terminus of the ilium and induce pernicious anemia, a macrocytic event. The pain and cramping inhibits appetite, causing weight loss. In some cases, the ulceration can be so severe that fistulas are formed and organs adjacent to the colon become affected, causing a host of potential secondary problems. The inflammation is often localized in the ilium (about 50% of the time) or at the junction of the ilium and the colon (about 30% of the time), and in the body of the colon about 20% of the time, but it can occur at any place, varying from the mouth the rectum. In some patients, the affected area is a continuum, whereas in others it develops simultaneously in different areas, creating a patchwork effect. Ulceration can even

happen outside the gastrointestinal system as, for example, in the mouth, skin, liver, joints, or eyes. What triggers the condition is unknown, but there appears to be a strong genetic component; approximately 20% of Crohn's disease patients have a first-degree relative who also is affected. The onset of the disease can start at any age but most typically appears between the ages of 15 and 35 years. It affects men and women to an equal extent, and Caucasians are at greater risk than other ethic groups.

(Choice 2) is the other inflammatory gastrointestinal disease. Unlike Crohn's disease ulcerative colitis causes inflammation only in the colon or rectum, almost always only involves the superficial layer of the inner lining of the bowel, and the inflammation generally is diffuse and uniform rather than concentrated in some areas and pitted and nonhomogeneous. However, some relation must exist between the two diseases since having first-degree relatives with ulcerative colitis is a risk factor for developing Crohn disease.

(Choice 3) is not a disease per se but a description of a group of functional disturbances that may cause symptoms such as bloating, flatulence, diarrhea and/or constipation, and mucus in the feces. Unlike Crohn disease or ulcerative colitis, the syndrome very rarely causes inflammation or any type of tissue damage. Irritable bowl syndrome is very common. There is no known cause, but individuals may have heightened sensitivity to certain foods or to stress. It is more common in young adults, particularly in females.

(Choice 4) is defined as too little of the intestinal enzyme lactase used to digest the lactose (milk sugar) found in the normal diet. By definition, all mammalian species drink milk at birth and therefore must produce sufficient amounts of lactase to thrive through infancy. However, as they age, most species develop an intolerance to lactose-containing foods (dairy products), and they lose the ability to synthesize sufficient lactase. This intolerance causes bloating and flatulence upon ingestion of dairy products. Western Caucasian Europeans and their descendents are the major exceptions to the rule that lactase synthesis ceases with age.

(Choice 5) There isn't a specific diagnosis or a recognized disease called "nervous stomach". Some doctors may use the term to generally describe symptoms of indigestion, bloating, or changes in bowel habits, especially after tests fail to reveal a specific cause.

18. Question **1 points**

A 27-year-old Irish Catholic woman gave birth to a 7- pound 5-ounce baby boy. The vaginal birthing was normal and everything seemed fine, but 26 days after the baby was born the mother began to have heavy vaginal bleeding. Her husband called 911, and an ambulance brought her to a nearby trauma facility. They managed to stop the bleeding, but she had lost so much blood that they gave her two units of replace- ment blood and admitted her into the hospital. There, the attending physician noticed she had many bruises on her legs, arms, and trunk, making him think abuse. But he took a careful history, starting with the questions: How did you get these bruises? Have you or any of your family ever had a bleeding problem before? She replied, "The bruises just appeared, but I always did bruise easily. As to bleeding problems I can't remember anything remarkable, except I used to have a bloody nose on a regular basis; so much so I some- times missed school; the school nurse told me to stop picking my nose, even though I didn't think I did." The physician also

asked: “How about your menses?” She replied, “No real problem, they occurred on a regular basis with minimal cramping, but I did lose a lot of blood. However, my family’s doctor told me not to worry about it since my mother and grandmother also had heavy bleeding during their periods, thus it must simply be a familial trait.” On the basis of this information, which of the following conditions does this lady most likely have?

1. Hemophilia A
2. Hemophilia B
3. Hemophilia C
4. Bernard-Soulier syndrome
5. von Willebrand disease ✔

INCORRECT ✖

The correct answer is 5.

Von Willebrand disease is the most common of all the bleeding disorders; it affects both sexes equally, with a prevalence accounting for somewhat over 1% of the population. There are three major forms of the disease, types 1, 2, and 3, with four subsets of type 2: namely, 2A, 2B, 2M, and 2N. Patients with type 1 have less than the normal amount of von Willebrand’s factor (vWP), patients with type 2 disease have aberrant forms of vWP, and patients with type 3 disease have very little if any vWP. Types 1 and 2 are inherited as dominant traits; type 1 is the most common form, accounting for about 70% of case, and type 2A is the next most common form, making up about 17% of the cases. The type 3 disease is inherited as a recessive trait, is the most severe form, and is also not commonly encountered. During pregnancy, the levels of vWP normally rise throughout the third trimester and then decrease, normally to baseline about a mouth postpartum. However, in patients with von Willebrand disease, the decrease is exaggerated and levels of vWP decline even below the pre-pregnancy levels, which were already less than optimal. Consequently, as in the case described, a hemolytic crisis may take place some weeks after the birth (usually about a month). Recent National, Heart, Lung and Blood Institute (NHLBI) guidelines state that health care providers should remain in close contact with women with von Willebrand disease during this period. Although von Willebrand disease affects males and females to an equal extent, females more commonly become aware of otherwise borderline cases because of their menstrual periods. As in the case described, it is not uncommon for generations of women to have abnormally heavy menstrual flow, and while true that it runs in families, that does not make it normal. The function of vWP is to bind to other factors involved in coagulation and bring them to wound sites, thus permitting them to work together to inhibit bleeding. The basic vWP subunit is a 2,050 amino acid protein with specific domains for coagulation factor VIII, heparin, collagen, and platelet receptors and hence platelets. These monomers are N-glycosylated and converted into multimers in a process involving cysteine cross-linking at the C-terminus. These multimers are very large, containing up to 80 monomers, and only these larger forms of vWP are functional. Pactor VIII

is rapidly degraded when it is not bound to vWP, thus low circulating levels of vWP are accompanied by low levels of factor VIII. As mentioned, type 1 von Willebrand disease is due to reduced levels of functional vWP. Remember that, since this is a dominant condition, affected individuals all should have at least 50% of the normal levels of functional factor inherited from the normal parent. Variation in activity then depends upon residual activity remaining in the aberrant factor inherited from the other parent; thus, type 1 patients have half to near normal vWP levels and activity. In type 3 von Willebrand disease, a child inherits an aberrant factor from both parents; consequently, the type 2 diseases involve lack of functionality or hyper-functionality in the factor that is synthesized.

(Choices 1 & 3) Hemophilia A and hemophilia B are both sex-linked recessive conditions and consequently are rarely expressed in females; thus, they are clearly incorrect choices. Hemophilia A is the most common form, with a prevalence of about 1 case per 5,000 male births and results in a deficiency of blood coagulation factor VIII; hence, an alternate name, factor VIII deficiency. Patients may have cases that vary from severe to minor bleeding problems. Hemophilia B results in a deficiency of coagulation factor IX, and is also called factor IX deficiency or Christmas disease, after the first patient in whom the disease was described. It is less common than hemophilia A, with a prevalence of about 1 case per 25,000 male births.

(Choice 3) causes a deficiency of coagulation factor XI and affects members of both sexes. It is inherited in a recessive fashion and is quite rare, affecting about 1 person among 100,000 adults, almost only people of Ashkenazi or Iraqi Jewish decent, making it unlikely that this Irish Catholic lady suffers from this disease.

(Choice 4) is a very rare condition, with an incidence of about 1 case per 1 million individuals; it is characterized by a thrombocytopenia. The biochemical cause is decreased expression of the glycoprotein complex Ib/IX/V on the surface of platelets. This complex normally serves as a vWP receptor, and when it is deficient, platelet adhesion to wound sites is inhibited. In vitro, this can be demonstrated by the lack of aggregation of platelets in response to ristocetin, an antibiotic that normally induces platelets to aggregate. A deficient number of platelets at the wound site inhibits formation of the primary platelet plug and causes an increased tendency to bleed. The thrombocytopenia may be due to decreased platelet half life.

19. Question **1 points**

A 26-year-old woman frantically presents to her physician showing him that she lost patches of scalp hair at various places. She screams, “I cannot continue living looking like this!” She blames her hair dresser, saying she left her under the dryer too long when she got a permanent and that she must have cooked her scalp. She is thinking of bring a lawsuit. She also says she expects the physician to provide treatment and to tell her if a suit is justified. Which of the following choices represents the physician’s best response? Tell her that:

1. He can perform a hair transplant that will restore her hair within a few months but she

doesn't have sufficient grounds for a suit.

2. He can perform a hair transplant that will restore her hair within a few months and he will testify on her account if she sues.

3. He feels confident that it is most likely that her hair will grow back spontaneously, but he has medication that may accelerate the process. However, there are no guarantees and he doesn't believe she has a case worth pursuing in court because the condition is an autoimmune reaction. ✔

4. He knows of several similar cases all caused by permanents and the end result always is complete baldness; so she should certainly sue.

5. He recommends shaving her head so the bald spots are no longer discernible; he will be glad to testify in court as to the extent of her loss.

INCORRECT ✖

The correct answer is 3.

This lady has a case of alopecia areata. Alopecia is a general term for baldness; alopecia areata is a common type of specific hair loss that causes coin-sized areas of baldness, usually on the scalp, although it may occur any place on the face or trunk. It affects 1%–2% of the population, both males and females, most often teens or young adults, but may occur at any age. Hair generally grows back spontaneously, and it is thought to be an autoimmune reaction; thus, **(Choice 3)** is correct and permanents are not suspected as a cause **(Choice 4)** is wrong). Person with this disorder sometimes have other autoimmune diseases, and there is a familial tendency to have the condition. As long as the loss is patchy and not all-inclusive, there is about an 80% chance it will grow back within a year. The early growth starts in the center of the bald patch; initially, it is fine and white, but with time, the hair usually regains its normal characteristics, although in older people it may not regain its color. The risk of not growing back is increased if there are large areas of hair loss, and in individuals with Down syndrome or eczema. If the hair loss on the scalp is total, resulting in baldness, the condition is called alopecia areata totalis; if the loss is all over the body, it is called alopecia areata universalis. These are relatively uncommon conditions accounting for less than 1% of all alopecia areata cases and in these cases regrowth usually does not happen.
(Choices 1 & 2) Alopecia areata usually does not lend itself to hair transplantation, therefore these **(Choices 1 & 2)** are incorrect. Some physicians would rather not treat alopecia areata, since odds are the hair will grow back spontaneously, but as it says in **(Choice 3)**, there are several treatments that may accelerate the rate of regrowth. Steroid
creams, steroid injections or pills, dithranol treatment, and physical irritation all have been used and seem to jump-start hair growth, but often hair loss starts again once treatment stops. Steroids in particular may induce serious side effects. The least disruptive and probably the most effective treatment is minoxidil lotion.
(Choice 5) Shaving her head is a counterproductive response, and is incorrect.

20. Question **1 points**

A 59-year-old man works as an accountant in a high- rise office building in Manhattan. Por many years, he habitually descended the 26 stories to the street at 10:30, noon, and 2:30 to smoke a cigarette and take a walk that lasted about a half hour during his lunch break. He figures walking is good for his health. However, starting about a month ago, he began to get pains in his calf that made walking difficult. These pains have become progressively worse, limiting his walks to just a few minutes before he had to find a place to sit and rest. He also found it hard to climb the stairs at home to go to bed; sometimes, he only managed three of four steps before he felt obliged to sit and rest. Worried, he consulted a physician who examined him and arrived at a tentative diagnosis. Which of the following choices best describes the first test the physician most likely preformed to verify this diagnosis?

1. Peripheral Doppler ultrasound
2. Peripheral angiography
3. Computed tomography (CT) scan
4. Ankle-brachial index (ABI) determination ✔
5. Magnetic resonance (MRI) scan

INCORRECT ✖

The correct answer is 4.

This man most likely has peripheral artery disease (PAD), sometimes called peripheral vascular disease (PVD), of the legs. The simplest and usually first diagnostic screening test is determination of the ankle-brachial index (ABI). This is done by simply determining the brachial and ankle systolic blood pressures and then dividing the brachial pressure into the value for the pressure at the ankle. A ratio greater than 0.9 is considered normal; a ratio between 0.8 and 0.9 is consistent with mild PAD, between 0.5 and 0.8 moderate PAD, and less than 0.5 severe disease. If the ABI is abnormal, a test (or tests) to better determine the site and quantify the extent of the lesion causing the occlusion may also be run.

(Choices 1,2 & 5) These imaging tests are likely to be peripheral Doppler ultrasound **(Choice 1)**, peripheral angiography **(Choice 2)**, or a computed tomography (CT) scan **(Choice 3)** would be non-contributory. Although a magnetic resonance imaging (MRI) scan **(Choice 5)** will provide an accurate image of the interior of the artery, it is less likely to be used because of the cost. In the United States, it is estimated that 10%-25% of the population over the age of 55 years has discernible PAD. Risk factors include smoking, diabetes, high blood pressure, dyslipidemia, obesity, male, African American descent, age older than 55, or having any other known vascular disease. Additionally, since the cause of PAD is most likely arteriosclerosis, having PAD suggests that a person is at risk of having other arterial diseases, such as coronary arteriosclerosis. Moreover, thrombi may break off from the arteriosclerotic regions in the leg and travel to the heart or lungs. The first step in treatment is smoking cessation. Subsequent steps are bringing other risks factors, such as diabetes or hypertension, under control. Accomplishing this is likely to require specific

medication in addition to lifestyle modification. Chelation therapy is a widely advertised alternative treatment; however, there is no sound evidence to support the use of this modality, which at the least can be expensive and on occasion dangerous. If such treatments are not beneficial, surgery (stents, bypass surgery and angioplasty, etc.) might be attempted. An unfortunate last resort most commonly utilized for diabetics is amputation.

21. Question **1 points**

A 29-year-old man visits his family physician with a history of pain in his legs. He states that the pain occurs in the evening and even at night, which results in disturbed sleep. He has even tried waking up and walking around to try to get rid of the pain. At times, he feels a tingling sensation in the legs, as if some- thing were crawling over them. Recently, he noticed a cramp like pain while he was driving, and he had to pull over. He had seen a psychiatrist recently and had been prescribed a neuroleptic medication. Based on the presentation, the most likely diagnosis to consider would be which of the following?

1. Painful legs and moving toes syndrome
2. Fibromyalgia
3. Restless leg syndrome ✔
4. Cauda equina syndrome
5. Akathisia

INCORRECT ✖

The correct answer is 3.

Restless leg syndrome is a disorder of unknown etiology that affects both men and women equally. These patients complain of disagreeable sensations in the legs, cramp like pains, sometimes a crawling sensation, or even itching. These patients try to move their limbs to get rid of the pain, even getting up to walk or pace around. While the pain tends to disappear with movement, it returns during the resting phase. The pain can disturb sleep. It usually peaks by midnight and thereafter tends to improve by morning. The problem can occur during the day, either while being sedentary or while driving a car. It may be associated with iron deficiency anemia, diabetes, pregnancy, and chronic renal failure, especially in patients who are on dialysis. Some patient may go for months without symptoms. There are no clinical tests, and the diagnosis is based on a history of dysesthesia, motor restlessness, a desire to move the limbs, and worsening of symptoms at night or during periods of rest. **(Choice 1)** does not occur at night and so does not induce insomnia. These patients complain of pain in the legs and feet, and involuntary writhing movements occur during this time. The pain does not get worse at the end of the day or during the night.

(Choice 2) usually involves a large area of the body, including the axial skeleton. Symptoms generally occur throughout the day, and movement does not improve it.
(Choice 4) is usually associated with low back pain, unilateral motor weakness, and sensory loss, and there may be loss of anal sphincter tone and problems with urination.
(Choice 5), an inner restlessness condition marked by an irresistible urge to move, is usually induced by medications such as phenothiazines or is a result of illness, such as Parkinson disease. These patients have restlessness that tends to affect the entire body, unlike restless leg syndrome, in which the problems are localized to the extremities. Symptoms occur less often at night, so insomnia is not a feature.

22. Question **1 points**

An 87-year-old man diagnosed with type 2 diabetes 27 years ago also has systolic hypertension and a body mass index (BMI) of 32. He presently is taking hydrochlorothiazide, atenolol, and amlodipine to control his systolic blood pressure, which when measured at home still averages about 167 mm Hg, and at the doctor's office, it ranges from 188 to 205 mm Hg. In addition, he also takes atorvastatin to help control his cholesterol level, which is now, with the help of this drug, at 125 mg/dL. In reviewing the results from a basic metabolic panel taken the previous week, his physician noted his serum creatinine level was 1.20 mg/dL (normal 0.70-1.30) and his blood urea nitrogen (BUN) 23 mg/dL (normal 9-23). After reviewing his history, the physician recommends starting enalapril to lower the patient's blood pressure. Three days after starting the enalapril, the patient checked himself into the emergency room (ER) complaining that he had hardly been able to urinate for the past 2 days. In addition, his arms and legs seemed to be swelling; he has abdominal pain, is unusually tired, and has problems concentrating. His serum creatinine level was now 1.80 mg/dL and his blood urea nitrogen (BUN) 36 mg/dL. Which of the following choices correctly describes the cause of this patient's symptoms?

1. Vasodilation of the postglomerular efferent arterioles ✔
2. Vasoconstriction of the preglomerular arterioles
3. Glomerulonephritis
4. Acute interstitial nephritis
5. A urethral stone

INCORRECT ✖

The correct answer is 1.

This man is suffering from acute renal failure (ARP) marked by oliguria and retention of nitrogenous wastes. Causes of ARP are customarily divided into three categories: prerenal, renal, and postrenal. Prerenal causes are those occurring in the blood supply leading up to

the glomerulus, renal are due to events in the body of the kidney, and postrenal are due to obstructions in the urinary tract leaving the kidney. In the case described, the precipitating factor was the ingestion of enalapril. As a general rule, angiotensin-converting enzyme (ACE) inhibitors such as enalapril or of angiotensin II receptor blockers (ARBs) are beneficial and lower morbidity and mortality. However, they can trigger ARP because they inhibit the production or utilization of angiotensin II, which will cause vasodilation of the postglomerular efferent arterioles; the result is a decrease of glomerular pressure, which decreases the filtration rate and the production of urine, leading to acute renal failure. This usually only happens in persons whose vascular system is stressed in some way. The man's system in this case was stressed by old age, diabetes, and evidence of "inflexible" arteries as demonstrated by his resistant systolic hypertension.

(Choice 2) Because the afferent preglomerular arterioles supply blood to the kidney, vasoconstriction of the preglomerular arterioles also may cause prerenal ARP. Non-steroid anti-inflammatory drugs (NSAIDs) may induce ARP via this mechanism because they inhibit cyclo-oxygenase (COX), and thus inhibit prostaglandin synthesis.

(Choice 3) rarely causes ARP, but it represents a type of renal ARP if it is quickly induced. It will be accompanied by systemic manifestations such as fever, rash, and arthritis.

(Choice 4) is another type of renal ARP. This condition often results from an allergic reaction to one of a host of drugs, including allopurinol, cephalosporins, cimetidine, ciprofloxacin, furosemide, NSAIDs, penicillins, phenytoin, rifampin, sulfonamides, and thiazide diuretics. Several diseases can also lead to development of acute interstitial nephritis. It again will be accompanied by a host of systemic manifestation. As indicated, obstruction of the outflow of urine from the kidney can also cause postrenal ARP.

(Choice 5) is a cause easily visualized. Other causes include prostatic hypertrophy or cancer, catheters, and strictures.

23. Question

1 points

A 54-year-old man presents to his physician for a physical examination. He is 5 ft 11 in tall and weighs 229 lb (body mass index [BMI] 33 kg/m^2). His blood pressure is 165/80 mm Hg, pulse is 75/min and regular. Otherwise, results of his physical examination are unremarkable. He was asked to have a fasting blood sample taken for a lipid profile. The next week, the following data are reported: total cholesterol, 225 mg/dL; triglyceride level, 200 mg/dL; low-density lipoprotein, 145 mg/dL; and high-density lipoprotein, 40 mg/dL. Because his BMI defines him as being class I obese and because his total cholesterol is borderline high, the physician puts him on a diet and prescribes a cholesterol-lowering medication. Since starting the medication, the patient experienced problems with constipation and a bloating sensation that is relieved by increasing dietary fiber. Which of the following metabolic alterations is expected from the drug he is taking?

1. Decreased activity of hydroxymethylglutaryl (HMG)-CoA reductase
2. Decreased synthesis of triglyceride in hepatocytes

3. Increased activity of capillary lipoprotein lipase
4. Increased excretion of free cholesterol in feces
5. Increased synthesis of hepatocyte low-density lipoprotein receptors ✔

INCORRECT ✖

The correct answer is 5.

The patient is most likely taking a bile acid-binding resin (e.g., cholestyramine) to lower his cholesterol level. These resins work by binding bile acids in the intestine to form an insoluble, nonabsorbable complex that is excreted in the feces along with the resin. This action reduces circulating cholesterol levels and causes a compensatory increase in the synthesis of hepatocyte low-density lipoprotein receptors to more efficiently recover reduced levels of cholesterol from the blood. Recovered cholesterol is then converted to bile acids to replenish the bile acids that are lost in the stool. The most common complaints are constipation and bloating that are relieved by increasing fiber or mixing psyllium seed with the resin. Heartburn and diarrhea are occasionally reported. Problems may also arise from the resin forming complexes and promoting the excretion of other substances such as digitalis, folic acid, thiazides, and warfarin.

(Choice 4) Normally, cholesterol is effectively reabsorbed from the intestine as part of the bile acids and is not excreted as the free compound. A bile acid-binding resin does not change this condition; however, now rather than being reabsorbed as an integral part of the bile acid, it is excreted as a component in a bile acid–bile acid binding resin complex, not as free cholesterol; increased excretion of free cholesterol in feces does not happen.

(Choices 1,2 & 3) The "statin" drugs (e.g., lovastatin) inhibit HMG-CoA reductase **(Choice 1)**, the rate-limiting enzyme of cholesterol synthesis. This decreases the synthesis of cholesterol. Rare side effects of statin drugs include hepatic toxicity with an increase in transaminases, and a myopathy with an increase in creatine kinase; however, evidence suggests that the statins are also anti-osteoporotic, an unexpected boon. Fibric acid derivatives (e.g., gemfibrozil) and nicotinic acid lower serum triglyceride (TG) levels. Fibric acid derivatives decrease hepatic TG synthesis **(Choice 2)** and also interfere with the formation of very-low-density lipoproteins (VLDLs) in the liver. Complications that might be associated with fibric acid derivatives include rash and myopathy, the latter sometimes causing rhabdomyolysis with marked elevation of creatine kinase and myoglobinuria. Nicotinic acid is also used in the treatment of hypertriglyceridemia (also hypercholesterolemia). The drug interferes with the formation of VLDL in the liver. Flushing is the major side effect of nicotinic acid and is diminished by taking a nonsteroidal anti-inflammatory drug 30 minutes before taking nicotinic acid. Flushing is not present in this patient. Fibric acid derivatives and nicotinic acid both activate capillary lipoprotein lipase **(Choice 3)**, promoting increased delivery of fatty acids to the adipose.

24. Question **1 points**

A 42-year-old man complains of headaches when he wakes up in the morning. He reveals that has had these headaches for maybe as long as a year, but they have been occurring more often and are getting progressively worse. He also feels as though he is losing his strength and that he sometimes has trouble placing his feet where he wants to without looking down at the ground. The physician observes him to have an unusually prominent jaw with spaces between the teeth, and large hands and feet. By asking him to press against his arm, the physician confirms muscle weakness. Upon physical examination, his blood pressure was found to be 135/95 mm Hg, and his non fasting blood glucose value was 195 mg/dL. Which of the following would be expected in this patient?

1. Chest radiograph with normal-sized heart
2. Normal-sized sella turcica
3. Lack of suppression of glucose with an oral glucose challenge ✔
4. Decreased concentration of insulin-like growth factor-1
5. Decreased serum growth hormone

INCORRECT ✖

The correct answer is 3.

The patient has acromegaly secondary to a benign pituitary adenoma with excess secretion of growth hormone {not decreased **(Choice 5)**} by the anterior pituitary and of insulin-like growth factor-1 {not decreased **(Choice 4)**} from the liver. If the condition occurs before fusion of the epiphysis, gigantism occurs.

Clinical findings associated with acromegaly include:

- Generalized enlargement of bone, cartilage, and soft tissue, resulting in large hands and feet, frontal bossing, a prominent jaw, an increase in hat size, spaces between the teeth, and hypertrophy of the left ventricle observed by chest radiography {not a normal-sized heart **(Choice 1)**. This hypertrophy may lead to cardiomegaly and cardiomyopathy with congestive heart failure (the most common cause of death).
- Diastolic hypertension (observed blood pressure is 135/95 mm Hg)
- Muscle weakness (his perception confirmed by the physician)
- Peripheral neuropathies (he can't be sure where his feet are, loss of proprioception)
- Diabetes mellitus from the gluconeogenic properties of growth hormone
- Headaches and visual field defects from encroachment on the optic chiasm

The laboratory findings include:

- Hyperglycemia (40%) (his serum glucose level is 195 mg/dL)
- Inability to suppress glucose with an oral glucose tolerance test **(Choice 3)**
- A paradoxical increase of serum growth hormone concentration with injection of thyrotropin releasing hormone (TRH)
- No stimulation of growth hormone release with L-dopa (normal persons have an increase in GH)

- Before fusion of the epiphysis occurs, increased serum phosphate associated with growth spurts
- Enlargement of the sella turcica in more than 90% of patients {not a normal-sized sella turcica **(Choice 2)**}

Treatment consists of transsphenoidal Surgery or the use of a somatostatin analogue called octreotide, which produces clinical improvement in 70% of cases.

25. Question **1 points**

For the past 6 months, a 55-year-old man with a 35year history of smoking cigarettes has complained of persistent frontal headaches. For several days, he has had projectile vomiting and bilateral blurry vision. Physical examination shows bilateral papilledema. A magnetic resonance imaging (MRI) study shows a mass lesion involving the corpus callosum that has spread to both cerebral hemispheres. The patient dies a few days later of a transtentorial herniation. Which of the following underlying conditions is the most likely cause of the lesion?

1. Cerebral infarction
2. Glioblastoma multiforme ✔
3. Intracerebral hemorrhage
4. Medulloblastoma
5. Metastatic lung carcinoma

INCORRECT ✖

The correct answer is 2.

The patient has a glioblastoma multiforme. This is the most common primary cancer of the brain in adults (peaks in the 40 to 70-year-old age bracket) and either arises de novo or from a preexisting low-grade astrocytoma. The tumors are located predominantly in the cerebral hemispheres and are classified as grade IV astrocytomas. There is no causal relationship with cigarette smoking. The distribution of this lesion and the history of headache, projectile vomiting, and bilateral papilledema are classic signs and symptoms of glioblastoma multiforme. The magnetic resonance imaging (MRI) scan shows a mass lesion extending across the splenium of the corpus callosum and into the adjacent cerebral hemisphere bilaterally. These tumors rarely metastasize out of the neuraxis. Radiation and chemotherapy are used for treatment. There is a 25%-40% 5-year survival rate.

(Choice 1) are most commonly caused by a thrombus overlying an atheromatous plaque, which causes a pale infarction that extends to the periphery of the cerebral cortex. The lesion in this patient does not adhere to this distribution.

(Choice 3) usually are due to hypertension (not recorded in this patient), which produces small-vessel aneurysms that result in rupture of the vessel and hemorrhage into the brain. The basal ganglia area is the most common site for the hemorrhage.
(Choice 4) usually are located in the midline of the cerebellum in children.
(Choice 5) Metastases to the brain usually are multifocal and occur primarily at the junction of the gray and white matter near the periphery of the brain.

26. Question **1 points**

An 18-year-old man enlisted in the Marines. He appeared to be in good physical shape, but his buddies sometimes remark about his unusual thirst, and he was annoyed by the fact that he often became so thirsty at night that he had to get up and drink a couple of glasses of water. Not surprisingly, he also had a need to urinate frequently. Since he acquired these traits around the age of 10, he did not regard them as being abnormal, just slightly annoying; his megathirst apparently started shortly after he fell from a tree and was knocked unconscious for a short time. He did fine during basic training until his unit went on a forced march during a hot August week. By the first night, he had emptied his canteen and suffered such a severe thirst that he actually woke up his sergeant begging for extra water. Luckily, his sergeant was a compassionate man and recognized something was truly wrong. In addition to his desperate pleading, he seemed too weak to stand straight at attention, looked and felt feverish, and his eyes appeared to have sunk deep into his forehead. Consequently, he was excused from the march and sent to the sickbay. Once there, his symptoms were recognized as dehydration, and he was given water and sent to the base hospital for further evaluation. The medics there quickly made a diagnosis and prescribed which of the following treatments?

1. Metformin
2. Foscarnet
3. Desmopressin ✔
4. Increasing his daily water intake
5. Demeclocycline

INCORRECT ✖

The correct answer is 3.

Clearly, this young man has a polydipsia/polyuria (excessive drinking/urinating) problem. Desmopressin is a vasopressin (also known as, the antidiuretic hormone) analog (1-deamino-8-D-arginine vasopressin) and is the only choice presented that would directly inhibit water loss into the urine. The word diabetes means to have an abnormal need to urinate. There are two basic types of diabetes, the more common “sweet” type, diabetes

mellitus, and the insipid, or “tasteless” type diabetes insipidus. Physicians presented with such a patient would order an electrolyte panel to help diagnose his condition, but even without additional data, common sense indicates this patient does not have diabetes mellitus {and therefore does not need metformin **(Choice 1)** since he has had this condition for many years and is healthy except for his abnormal thirst; it follows he must have diabetes insipidus. However, there are five recognized types of diabetes insipidus:

1. Central (also known as, neurogenic or pituitary), in which the hypothalamus has lost the capacity to synthesize vasopressin; this condition may be idiopathic or due to some sort of injury, physical or pathological (tumors or infection).
2. Nephrogenic, in which the kidney tubules have lost functional binding sites for vasopressin; this may be caused by kidney disease, drugs that inhibit binding, or in very rare cases, it is inherited.
3. Dipsogenic, in which the thirst-regulating part of the brain is damaged.
4. Gestational, caused by the placenta’s destruction of vasopressin. As a rule, recovery is spontaneous within 24 days after birth, although an affected woman is at high risk to develop another case of diabetes insipidus during subsequent pregnancies.
5. Psychogenic, in which a patient has an obsessive compulsion to drink large quantities of water.

Usually, a physician will measure blood glucose, bicarbonate, and electrolyte levels, as well as urinary osmolarity, as the first step in confirming diabetes insipidus. Typically, urine osmolarity and electrolyte levels are lower than normal and the degree to which hypernatremia occurs reflects the degree of dehydration. A fluid deprivation test helps determine if the diabetes is due to lack of vasopressin (central), to the kidney’s insensitivity to vasopressin (nephrogenic), or to excessive fluid ingestion (dipso or psychogenic). Normally, as fluid deprivation proceeds, dehydration starts to occur and the kidney compensates by excreting a lesser volume of more concentrated urine; consequently, the body retains a greater volume of water. Patients with diabetes insipidus fail to concentrate their urine; however, patients having central diabetes insipidus will start to concentrate their urine after the administration of desmopressin. This will not occur if the patient has nephrogenic, dipsogenic, or psychogenic diabetic insipidus.

(Choices 2 & 4) Foscarnet, an antibiotic used to treat cytomegalovirus infection, has recently been shown to cause nephrogenic diabetes insipidus in certain patients and consequently will not likely be used to treat any form of diabetes insipidus. Increasing his daily water intake **(Choice 4)** also would be counterproductive and might lead to water intoxication caused by hyponatremia.

(Choice 5) is a broad-spectrum tetracycline antibiotic and the only tetracycline with the property of competing with vasopressin for its binding sites on the renal tubules, thus it too may cause a transient diabetes insipidus and will not likely be used to treat diabetes insipidus.

27. Question **1 points**

While on a camping trip with his son in a remote location in Alaska a 69-year-old man accidently got a fish hook imbedded in the palm of his hand. They dug out the hook, washed the wound, and bandaged it. Apparently, they did not clean it out adequately because when they got back home a week later, it showed signs of infection. A doctor at the local ER cleaned it out thoroughly and gave him a shot of penicillin. The wound healed nicely but several weeks later, the man developed a mild fever, which persisted in the absence of any other symptoms. After a month, his temperature suddenly peaked and he developed excessive sweating, chills, fatigue, muscle and joint pains, night sweats, and painful lesions on the pads of the fingers. Which of the following is the most likely cause of this man's symptoms?

1. Infection by Staphylococcus aureus ✔
2. Infection by Streptococcus viridans
3. Infection by Candida albicans
4. Infection by Streptococcus bovis
5. Infection by Clostridium septicum

INCORRECT ✖

The correct answer is 1.

Bacteria that infected this man's wound multiplied, entered his circulatory system, and localized in one of his heart valves. Initially, this caused a case of subacute endocarditis marked by a low-grade fever. The infection then fulminated, causing symptoms commonly associated with acute infectious endocarditis, including a higher temperature, excessive sweating, chills, fatigue, muscle and joint pains, night sweats, and painful lesions, called Osler's nodes, on the pads of the fingers. Infections initiated by breaks in the skin are often caused by Staphylococcus aureus. In this case, because the shot of penicillin received at the ER did not prevent the development of endocarditis, it is likely that this was a methicillin-resistant strain. Unless faced with a massive infusion of organisms, skin breaks as a rule do not cause endocarditis because healthy valves are smooth and don't provide a surface for organisms to adhere to. However, in older individuals, one or more valves are sometimes affected by arthrosclerosis and become roughened by calcification; this provides a place for organisms to anchor and proliferate, as presumably happened in this gentleman. This is the reason why the incidence of endocarditis is higher in the elderly.

(Choice 2) Streptococcus viridans is an organism found in the mouth and is associated with endocarditis after dental work. Prior to October 2007, dentists were advised to administer penicillin to patients at risk for heart infection as prophylaxis to prevent endocarditis. However, at that time, the American Heart Association published an article in Circulation recommending that most of these patients no longer get short-term antibiotics as a preventive measure before their dental treatment. The consensus was that the risks of taking preventive antibiotics outweigh the benefits for most patients. These risks include adverse

reactions to antibiotics that range from mild to potentially severe and, in very rare cases, death. Inappropriate use of antibiotics can also lead to the development of drug-resistant bacteria.
(Choice 3) Infection by the yeast Candida albicans most commonly occurs in IV drug users; these infections commonly enter the right heart and often infect the tricuspid valve.
(Choices 4 & 5) Streptococcus bovis and Clostridium septicum are part of the natural flora of the colon and cause endocarditis if the bowel is pierced or in colon cancers, rarely from skin punctures.

28. Question **1 points**

An internist at a major medical center received a letter from a physician located far away from his place. The letter said: “With your permission, I am asking a former patient of mine to see you. He is an 18-year-old man suffering from occipital bone syndrome. Physically, he has the pathognomonic bilateral exostosis of the skull and pili torti, extremely loose skin and joints, long neck, dysautonomia, and vascular tortuosity, as well as a somewhat unusual appearance including an abnormally long neck and face, with a high forehead and an arched palate. He also suffers from seizures, which are presently under control by medication. He has a very low normal IQ of 88. I am sure you will find his case interesting and his continued treatment challenging. If you accept him as your patient, I will send you his medical records.” The patient described in this letter has an aberration in the metabolism of which one of the following trace elements?

1. Zinc
2. Selenium
3. Chromium
4. Copper ✔
5. Cobalt
6. Manganese

INCORRECT ✖

The correct answer is 4.

There are three inborn errors of copper metabolism: Wilson disease, Menkes disease, and occipital horn syndrome. Both occipital horn syndrome (once known as Ehlers-Danlos syndrome IX and sometimes called X-linked cutis laxa) and Menkes syndrome (also known as, kinky hair syndrome) are caused by mutations in ATP7A located on Xq12-13. The normal gene has 23 exons and encodes a P-type copper transport protein with ATPase activity. Mutations in the splicing sites may result in insertions or deletions and nonsense rearrangements. Such errors result in greater distortion of the protein product in patients

having Menkes disease than they do in patients with occipital horn syndrome; thus, boys afflicted with Menkes disease are by definition sicker than those with occipital horn syndrome.
Male Menkes babies tend to be born a little prematurely; they seem normal at first, but then spiral into a tragic decline. Usually by about the third month, the child has an epileptic event and his appearance takes on the typical manifestation of the disease, which includes light, often steely colored, kinky hair that is sparse on the sides, plus an unusual appearance. Prom that time on, the child fails to thrive and degenerates neurologically and physically, eventually dying, generally by the third year. The normal Menkes gene product exists in both a long and short form, with different tissue distribution. The shorter form is expressed in occipital horn syndrome, and this probably accounts for the milder phenotype, which is similar to the one described in the case history. Formerly, most afflicted boys died in their early teens but now commonly live well into their 20s.
The normal Menkes and Wilson disease genes have about 55% identity, and both code for copper-transporting proteins, but their distribution and the function of their gene products are different. The underlying function of the normal Menkes gene product is to bring copper ion into the body and distribute it among cells; in contrast, the function of the normal Wilson gene product is to remove excess copper ion from the body. Consequently, the normal Wilson disease gene is expressed predominantly in the liver, whereas the normal Menkes disease gene is not expressed to a significant extent in liver but is in most other tissues; moreover, serum copper ion levels are lower than normal in both Menkes disease and occipital horn syndrome, but raised in Wilson disease.
(Choice 1) A rare autosomal recessive condition called acrodermatitis enteropathica (also known as, Brandt syndrome, Danbolt-Cross syndrome or congenital zinc deficiency) causes a congenital form of zinc deficiency. Zinc and copper are the only trace elements associated with known inborn errors of metabolism. Zinc is a cofactor for as many as 300 enzymatic reactions; hence, a deficiency can have profound effects. Patients having acrodermatitis enteropathica who are not treated by lifelong dietary zinc supplementation die within a few years. Acquired deficiencies are more common; the deficiency may be due to dietary insufficiency (either because of poor nutrition or to conditions such as anorexia nervosa or alcohol abuse), a decreased ability to absorb the ion (either because of intestinal disorders or sometimes associated with aging), and in patients on complete parenteral nutrition, insufficient zinc allowance. Deficiency states in developed counties are generally modest and the effects subtle. An obvious effect is in limiting the growth of children. Earlier studies in several U.S. communities showed that dietary supplementation with zinc induced growth spurts, thus indicating that these children had a nutritional deficiency. Dermatologic changes (e.g., rashes) are another readily recognized consequence of zinc deficiency. Loss of acuity of taste (dysgeusia) or smell (anosmia) caused by a reduced ability to absorb zinc has been observed among aged individuals. Chronic zinc deficiency also increases susceptibility to infection by reducing immunologic efficiency and also by reducing the efficiency of wound healing because it is a cofactor for collagenase, which is required to Resorb type III collagen so that it may be replaced by type I collagen. As a rule, these acquired deficiencies can be reversed by zinc supplementation, but with a note of caution: taking large quantities of zinc can reduce the bioavailability of copper because zinc induces the synthesis of a metallo thioneine that binds copper within intestinal cells and prevents its absorption. Thus, individuals taking zinc supplements are advised to also take a copper supplement.

(Choice 2) is a cofactor for glutathione peroxidase (which neutralizes hydrogen peroxide and peroxide free radicals in cells), thioredoxin reductase (which indirectly reduces certain oxidized molecules), and three deiodinase enzymes (which interconvert forms of thyroid hormone). Its glutathione peroxidase activity helps reduce free radicals and is thought to act to reduce the risk of cancer. A frank selenium deficiency also predisposes patients to Keshan disease, an endemic viral cardiomyopathy primarily affecting children and young women. At high doses, selenium is toxic, causing hair loss, abnormal nails, dermatitis, peripheral neuropathy, nausea, diarrhea, fatigue, irritability, and a garlicky breath odor.
(Choice 3) is another essential trace element required by humans. It is almost ubiquitous in nature, making deficiency states essentially unknown. Promotional literature from supplement distributors may claim chromium supplements can reduce serum cholesterol levels, prevent or help alleviate symptoms associated with diabetes, reduce body fat, and help build muscle, but there is little evidence to substantiate such claims. Nonetheless, chromium is a component of glucose tolerance factor, and consequently does help insulin make glucose available to cells by facilitating the binding of insulin to its receptors located in adipose and muscle. Thus, if a deficiency of chromium truly did exist, it might be associated with impaired glucose tolerance.
(Choice 5) The human requirement for cobalt is not for the ionic form of the metal, but for a preformed metallo vitamin that cannot be synthesized from dietary metal. Therefore, it is the vitamin B12 content of foods in the diet that is of importance in human nutrition.
(Choice 6) is a cofactor for arginase, pyruvate carboxylase, glutamine synthetase, and manganese superoxide dismutase (SOD), and is the preferred metal cofactor for glycosyltransferases. No deficiency states for manganese have been established.

29. Question **1 points**

A poorly dressed 56-year-old lady enters an emergency department facility at a local hospital in an agitated manner saying she needs attention immediately because she just discovered she has colon cancer. The triage nurse calms her down a bit by asking her what makes her think that is true. She responds: “For the past week, each time I make number two, I get a sharp pain, and because the pain lasts for around a half hour after I wipe myself I know it must be coming from my insides. The worst was today when I saw bright red blood; I heard on the TV that blood when you go to the bathroom is a sign of cancer.” The nurse tells her that there are also many other additional causes for blood in the stool. She finds that the lady is hypertensive but has not seen a physician since she was pregnant at the age of 18 years; the nurse then arranges for the woman to see one of the ER doctors. When the doctor sees the patient, he arranges for her to see a primary care physician but also recommends treatment. Which of the following choices is the most probable treatment recommended?

1. Removal of a mass that may be protruding from the patient’s anus
2. Perform a sigmoidoscopy and during this procedure excise any polyps found

3. Application of a local steroid cream twice daily, plus use of stool softeners and hot sitz baths ✔

4. Prescription of antihypertensive drugs and antibiotics

5. Cautery by argon plasma coagulation applied via an endoscope

INCORRECT ✖

The correct answer is 3.

This lady most likely suffers from an anal fissure, for which the classical treatment is application of a local steroid cream twice daily, plus use of stool softeners and hot sitz baths. Such a fissure results from the passage of a large, hard bolus of stool and is a common cause of rectal bleeding. Severe pain is an important symptom, and the condition is confirmed by observation of a semi-epileptical defect in the anal skin running in a radial direction. The treatment described usually results in complete recovery.

(Choice 1) A mass protruding from the patient's anus is usually due to a prolapsed internal hemorrhoid and generally is caused by straining to defecate. The mass generally retracts spontaneously and is rarely accompanied by pain unless it is thrombosed.

(Choice 2) There is no call for a sigmoidoscopy, and polyps cannot be removed during a sigmoidoscopy.

(Choice 4) Prescription of antihypertensive drugs and antibiotics is a therapy used to treat relatively mild cases of ischemic colitis. Control of hypertension will facilitate blood flow into the colon, while the antibiotics will help prevent infection.

(Choice 5) Idiopathic bleeding from the bowel due to dilated blood vessels in the mucosa is called angiodysplasia and may be initially treated by cautery employing argon plasma coagulation applied via an endoscope.

30. Question **1 points**

A 79-year-old man with a long history of systolic hypertension was enrolled in a physical therapy session because of chronic back pain. For a reason that was unknown at the time, it was becoming increasing difficult for him to perform exercises involving raising his legs. As a result, the therapist decided to massage his legs and noted they were distended with edema. Consequently, he made an appointment with his primary care physician, who performed an electrocardiogram (ECG); this was essentially normal. The physician also prescribed a diuretic and several tests, and referred him to a cardiologist. Which one of the following tests will best provide quantitative evidence defining the severity of the problem suspected by the primary care physician?

1. Atrial natriuretic peptide (ANP)

2. Brain type (B-type) natriuretic peptide (BNP) ✔

3. C-type natriuretic peptide

4. A chest x-ray

5. A complete blood count (CBC)

INCORRECT ✖

The correct answer is 2.

The onset of edema is a possible symptom of congestive heart failure, which in turn may be caused by longterm hypertension and/or arthrosclerosis. This probable diagnosis needs to be confirmed, as must the degree of the problem. Among the tests outlined, this can best be done by determining the level of brain type (B-type) natriuretic peptide (BNP) in the serum. The natriuretic peptides are a family of polypeptides that cause the excretion of sodium into the urine; that is, they promote natriuresis. The different forms of the natriuretic peptides are named by the tissues from which they were first isolated. Even though BNP was first isolated from brain, its medical significance lies in the fact that this peptide is released from the heart's ventricles in response to excess stretching of the ventricular myocytes as often occurs in the stressed heart in congestive heart failure. In addition to stimulating natriuresis, this peptide also decreases systemic vascular resistance, thus causing a decrease in central vascular pressure, inducing and permitting a decrease in cardiac output.

At first, it was thought that measuring the serum level of BNP would provide a quantitative estimate of the degree to which the heart is damaged, but at best it is semiquantitative and conditions other than congestive heart failure may also increase the activity. The most definitive test of heart function and thus the best test to estimate the extent to which the heart is damaged is a stress echocardiogram. However, the measurement of the serum level of the BNP can be done in the doctor's office or at the bedside while an echocardiogram, in particular a stress echocardiogram, takes special equipment that is not always readily available, particularly in a primary care physician's office or in remote areas and Third World countries. In addition to using BNP as a diagnostic aid, a recombinant human BNP, nesiritide, has been approved for use in the acute treatment of decompensated congestive heart failure caused by systolic dysfunction. Unfortunately, it must be administered IV and has a very short half-life.

(Choice 1) acts similarly to BNP but is released from the atrium. However, its half-life in the serum is much shorter than that of BNP, which limits its usefulness as a diagnostic tool.

(Choice 3) is also found in brain and may play a role similar to BNP but its medical value has not yet been established.

(Choice 4) can show an enlarged heart but that in itself is not diagnostic of congestive heart failure nor does it provide even semiquantitative information concerning severity.

(Choice 5) is irrelevant.

31. Question **1 points**

A 48-year-old patient presents to the emergency room with a history of tiredness, vomiting, and abdominal pain. He has a long history of hypertension, diabetes mellitus, and hyperlipidemia, and during this past year, he developed end-stage renal disease. He admits to being depressed of late and has not kept his appointment with the nephrologist for renal dialysis, which he used to go to three times a week. The patient is on medications for hypertension, diabetes, and hyperlipidemia, in addition to medications for renal failure. Which of the following is the initial medication that should be administered to reverse the clinical condition reflected by the accompanying electrocardiogram pattern?

(https://www.amcquestionbank.com/wp-content/uploads/2015/11/Picture-Test-6-Q08.png)

1. Sodium bicarbonate
2. Furosemide
3. Calcium gluconate ✔
4. Glucose plus insulin
5. Cation-exchange resin

INCORRECT ✖

The correct answer is 3.

The electrocardiogram (ECG) pattern shown indicates that this patient has hyperkalemia. It reveals tall, slender, tented T waves in leads I, II, aVF, and V2 through V6. Other findings suggesting hyperkalemia include a widened QRS complex and even biphasic QRS-T complexes. Although hyperkalemia produces these characteristic patterns, the ECG is not a sensitive indicator of hyperkalemia, as almost 50% of patients with serum potassium levels above 6.5 mEq/L do not show any changes in the ECG pattern. This is because atrial cells are more sensitive than ventricular cells to elevated levels of potassium, permitting normal conduction even if atrial depolarization is inhibited; as a result, a junctional rhythm can result. To avoid dangerous arrhythmias and asystole, hyperkalemia above 6.5 mEq/L warrants immediate correction. In this case, hyperkalemia also is suspected clinically because the patient has end-stage renal disease, has not been undergoing dialysis, and has a history of tiredness, vomiting, and abdominal pain. Hyperkalemia could, in addition, have caused motor paralysis. Calcium gluconate 10% or 5% calcium chloride is the initial drug of choice. Approximately 5-30 mL of either administered intravenously provides calcium ion, which acts within minutes to antagonize cardiac conduction abnormalities; the effect lasts approximately an hour. In patients who are on digoxin, one must ensure that they do not have digoxin toxicity; if that were the case, calcium would only increase its toxic effects on the myocardium.

(Choice 4) Although administering calcium has a rapid effect, it does not promote the shift of potassium into the cells; consequently, its effect is transitory. To secure a more permanent effect, one usually follows up with insulin and glucose. This drives potassium into the cells,

thereby lowering serum potassium levels. Regular insulin (5-10 units) combined with 25 g of 50% glucose is administered intravenously. Insulin takes about 15 minutes to an hour to act, and the action lasts anywhere from 4 to 6 hours.
(Choice 1) Use of sodium bicarbonate is an alternative to insulin plus glucose. It creates a metabolic alkalosis, causing potassium ion to enter the cells in exchange for hydrogen ions. One or two ampules of sodium bicarbonate (approximately 44 mEq each) can be administered intravenously. Its effects begin in approximately 15-30 minutes and last anywhere from 1 to 2 hours. Thus, it is not as long lasting as insulin.
(Choice 2) The use of furosemide is inappropriate in an emergency situation because it takes anywhere from 30 minutes to 2 hours to lower potassium levels, and one may very well not have the luxury of time. It is a loop diuretic that exchanges sodium for potassium, expelling the latter in the urine.
(Choice 5) Like loop diuretics, sodium polystyrene sulfonate in 20% sorbitol, a cation exchange resin, may be used in nonemergency situations; 15-30 mL is administered orally or rectally. Potassium ions bind to the resin, lowering serum potassium levels. Due to the mode of administration, it takes much longer to act; its action lasts approximately 3 hours.

32. Question **1 points**

A 40-year-old man presents to a physician complaining of erectile dysfunction. In taking a history, it is determined that he started drinking alcohol at the age of 14 years and has continued since. Presently, he starts his day by having a beer or two with his breakfast, and he carries a flask of whiskey with him to help him get thorough the day. In the evening, he often polishes off a pint of whiskey before going to bed. However, he also states that he has a “hollow leg,” and the alcohol doesn’t affect his functioning in any way. Physical examination reveals a distended abdomen and dependent pitting edema. He also has numerous radially oriented vessels around a central core on his face, neck, and upper trunk. Which additional physical finding in this patient has the same pathogenesis as the skin lesion?

1. Ascites
2. Asterixis
3. Caput medusae
4. Esophageal varices
5. Gynecomastia ✔

INCORRECT ✖

The correct answer is 5.
The patient has cirrhosis of the liver (history of alcohol abuse, ascites, and dependent pitting edema). The skin lesions are spider angiomas, which have a central spiral arteriole with a group of small vessels radiating from the arteriole. Spider angiomas are associated with hyperestrinism, which is a complication of cirrhosis. In cirrhosis, the dysfunctional liver is unable to metabolize estrogen, which produces hyperestrinism. Hyperestrinism in men causes gynecomastia (development of breast tissue in males) and female secondary sex characteristics (palmar erythema, soft skin, and female hair distribution).
(Choice 1) Although the distended abdomen is due to ascites, the question asks for additional symptoms having the same pathogenesis as the skin lesion. The factors that contribute to the development of ascites include portal hypertension (increase in hydrostatic pressure), hypoalbuminemia (decrease in oncotic pressure), secondary aldosteronism (salt retention), and increased lymphatic drainage into the peritoneal cavity.
(Choice 2), or flapping tremor, refers to the inability to sustain posture. It is a sign of hepatic encephalopathy, which is caused by an increase in ammonia and false neurotransmitters (e.g., -aminobenzoic acid).
(Choice 3) are dilated periumbilical veins that are associated with increased venous pressure caused by portal hypertension.
(Choice 4) are dilated left gastric coronary veins, which normally drain the distal esophagus and proximal stomach and empty into the portal vein. An increase in portal vein pressure leads to dilation of the gastric veins (varices), which commonly rupture.

33. Question **1 points**

A 23-year-old male prelaw student presents to the ER with the complaint of an itch and rash in his genital region. In providing a history, he states that the itching is worse at night, making avoidance of scratching almost impossible. He adds that the morning after his first night of discomfort, he noted a few small red blisters and bumps on his penis and while trying to treat the condition he bathed and applied hand lotion. The next night, the itching was even more intense, and in the morning, he noted the head of his penis was fully involved and the rash had spread to the surrounding genital area. This induced him to seek medical help. The attending physician asks if the student had had sex within the past 4–8 weeks? The student tells him that he did have sex with a young woman he met at a bar about a month ago; however, he felt he was protected since he wore a condom. The attending physician replies, “I am 99% sure I know what ails you, and I am going to write a prescription. If I am correct and you follow the directions, your condition should clear up within a few days. If it doesn’t, let me know; otherwise, there should be no problem. I would also like to talk to the young lady if that is possible.” Which of the following choices most likely describes the prescription given?

1. Topical permethrin 5% ✔
2. Oral azithromycin

3. Oral ivermectin
4. Topical lindane (-benzene hexachloride) 1%
5. Parental penicillin G

INCORRECT ✖

The correct answer is 1.

The patient suffers from a case of scabies caused by a small, almost microscopic (0.3–0.9 mm) mite called Sarcoptes scabei, variety hominis. The recommended treatment for adults is 5% permethrin lotion, applied on a clean body from the neck to the toes and left for at least 8 hours, after which it can be washed off. A second treatment is recommended a week later. To ensure that all eggs and mites are killed, all clothing, bedding, towels, etc. used by the infected person should be washed in hot water and dried in a hot dryer before treatment begins. No special cleaning is needed for rugs, floors, coats, and furniture because the mites cannot survive away from a human body. When patients are infested, pregnant female mites burrow into the stratum corneum and deposit eggs. In 3–10 days, the eggs hatch into larvae, which then molt and mature into a new generation of adults via a nymphal stage. The eggs produce an allergic reaction that is intensified by the action of the maturing mites; this induces the rash and the itch. A first-time infection may take as long as 8 weeks for the full-blown allergic reaction to develop. On the other hand, a person sensitized by a previous infection will develop symptoms much more rapidly. The mites, and consequently scabies, are transmitted readily by skin-to-skin contact with an infected person. Thus, clusters of cases often occur within a household or a health care facility. Obviously, sexual intercourse involves skin-to-skin contact, thus causing scabies to be classified as a sexually transmitted disease. It is quite possible that this patient's partner picked up a few mites while working as a nurse and has already shown signs of scabies on her hands, a common site for early scabies to appear. Perhaps she only recognizes it as unusually red and rough hands and is spreading the disease among other patients; hence, the value of informing her that she is a vector. An epidemic of scabies in a health care facility can create havoc. Absolute diagnosis depends upon isolation and microscopic identification of active mites. However, it is more practical to make a tentative diagnosis based upon appearance and confirmed by response to treatment. Treatment as described above results in a 100% cure rate

(Choice 2) is an antibiotic commonly used to treated chlamydial infections. Scratching scabies infection can sometimes lead to thick scaling, particularly in immunocompromised persons. Such a condition is called Norwegian scabies, and the extensive scaling makes topical medication ineffective.

(Choice 3) Consequently, Norwegian scabies is treated with ivermectin, a broad-spectrum antiparasitic medication.

(Choice 4) is an organochlorine insecticide once used as the primary treatment for scabies. It functions as a neurotoxin by interacting with -aminobutyric acid (GABA) A receptor. In humans, it has a dermal LD50 of 1,000 mg/kg and has also been reported to be a carcinogen in rodents. Because of potential toxicity, it is now banned in 50 countries and approved by the U.S. Food and Drug Administration only as a second-line treatment for scabies or for pubic (crabs) and head lice.

(Choice 5) is an antibiotic often used to treat primary syphilis infections but has no role in treating primary scabies infections.

34. Question

1 points

For approximately the past two decades, a 62-yearold man had been a successful office manager at a new car dealership; for most of this time, he had his first alcoholic drink of the day upon arising and one or two more before arriving at work, then easily finishing off another pint of bourbon after leaving work and before going to bed. He felt alcoholic drinks had little effect on his ability to function and bragged about having a “hollow leg.” Until recently, he had never had anything to drink while at work, and despite his extreme drinking habits, his work had not suffered. Lately, however, he has had an irresistible urge to indulge in a drink or two or more, even during the workday, and he began to exhibit symptoms of impaired performance. Moreover, he had been caught telling untruths to cover up shortcomings in his performance. Consequently, the owner of the dealership threatened to fire him unless he admitted himself into an alcohol and drug rehabilitation center. As part of his admission to such a center, he underwent a thorough physical examination during which an abnormality present in most heavy drinkers one that may be reversed by abstinence was uncovered. This abnormality most likely was which one of the following?

1. Alcoholic hepatitis
2. Liver cirrhosis
3. Portal hypertension
4. An increase in the serum alanine aminotransferase (ALT) activity greater than an increase in the serum aspartate aminotransferase (AST) activity
5. Wernicke disease (Wernicke’s encephalopathy)
6. Korsakoff psychosis (Korsakoff syndrome)
7. Fatty liver ✔

INCORRECT ✖

The correct answer is 7.

Alcohol abuse and alcoholism are major problems in the Australia, estimated to affect over 8% of the adult population. Alcoholism, also known as alcohol dependence, is a disease that includes an almost irresistible craving for a drink; an inability to stop drinking once started; physical dependence marked by withdrawal symptoms such as nausea, sweating, shaking, and profound anxiety once drinking is stopped; and, a requirement to drink greater amounts to get a high as time progress, a phenomena called tolerance. Alcohol abuse is defined as drinking alcohol to such a degree that it causes personal harm, socially, physically, or with

the law as in accumulating drinking under the influence of alcohol (DUIs); however, in contrast to true alcoholism, physical dependence is not a characteristic. Whether a person is afflicted with alcoholism or abuse, drinking large quantities of alcohol causes many different pathological changes in the drinker's body. These abnormalities probably affect all heavy drinkers, although there is great variability regarding individual responses; thus, the pattern of symptoms shown varies among different individuals. Fatty liver is an abnormality found to be present in almost all heavy drinkers; fortunately, it frequently can be reversed by abstinence. Although fatty liver itself may not cause permanent damage, it is a symptom of abuse and a strong warning indicating a person should stop drinking before permanent damage is done. Generally, it is indicated by a slightly enlarged liver accompanied by higher than normal activity levels of liver enzymes in the serum. A computed tomography scan will show a liver that is less dense than normal, whereas an ultrasound test will produce a bright image in a ripple pattern. However, an absolute diagnosis requires a biopsy.

(Choices 1 & 2) Up to 35% of chronic drinkers develop an inflammation of the liver know as alcoholic hepatis. This is caused by the oxidative metabolism of alcohol by the liver that produces free radicals as a side product and also by reaction to acetaldehyde, a toxic intermediate. Alcohol also permits the passage of bacterial endotoxins from the intestine to the small intestine via the portal vein. These toxins further damage liver cells causing the release of inflammation-promoting cytokines, which set up a vicious cycle by stimulating the release of additional cytokines. These inflammatory processes deplete oxygen levels, inducing cell death, and cause fibrous scarring, a condition known as cirrhosis. Early hepatis may be reversed by abstinence, but cirrhosis is not reversible and in 10%–20% of drinkers, these inflammatory processes lead to cirrhosis severe enough to eventually cause death if the patient is not given a liver transplant.

(Choice 3) The fibrous scar tissue of cirrhosis also blocks the flow of blood through the branches of the portal vein that traverses the liver. This causes portal hypertension; when the portal pressure becomes high enough, blood is forced into other veins en route to the heart. These include veins in the esophagus, causing esophageal varices, and veins in the skin over the abdomen, leading to the appearance of spider veins. Esophageal varices are prone to bleeding, which may result in an anemia and may contribute to eventual death. Liver disease is almost always associated with leakage of enzymes from affected liver cells, causing an increase of these enzymes in the serum. Two of the liver enzymes are serum alanine aminotransferase (ALT) and serum aspartate aminotransferase (AST); however, in cases involving alcohol abuse, the increase in AST activity is always greater than that of ALT; thus **(Choice 4)** is incorrect.

(Choice 5) Chronic excess alcohol consumption of course damages additional organs, the brain being one of the more critical. Wernicke disease affects nerves in both the central and peripheral nervous systems. The underlying cause is believed to be malnutrition, primarily a lack of thiamine. Heavy drinking is often associated with a poor diet, but individuals who maintain a balanced diet may still suffer from a lack of thiamine because alcohol inhibits thiamine absorption from the gut. Symptoms associated with Wernicke disease include confusion, nystagmus, ophthalmoplegia, anisocoria, ataxia, poor pupillary reflexes, and in extreme cases, coma and death.

(Choice 6) often follows Wernicke disease, and although it too may involve thiamine deficiency, Korsakoff psychosis is associated with a direct toxic effect of alcohol on the brain that causes general cerebral deterioration, particularly affecting the medial thalamus and the

mammary bodies of the hippocampus. The six major symptoms of Korsakoff 's psychosis are blackouts after a bout of drinking, known as anterograde amnesia; loss of memory of people and/or effects occurring prior to a bout of drinking, known as retrograde amnesia (this symptom often follows delirium tremens); invention of memories to fill in gaps associated with blackouts or other losses of memory, known as confabulation (confabulation may not be purposeful, and the false accounts are often believed by the teller); evasion of meaningful conversation; loss of insight; and increased apathy. These changes involve loss of neural function and bleeding within the mammary bodies. Although some improvement may occur during abstinence and treatment with thiamine supplementation, the condition cannot be reversed. It has been hypothesized that individuals genetically susceptible to symptoms associated with thiamine deficiency are more susceptible to acquiring the Wernicke and Korsakoff conditions.

35. Question

1 points

A 30-year-old man with a history of chronic diarrhea describes his stools as greasy and foul smelling. He recently developed pruritic vesicular lesions involving the elbows. A quantitative stool test for fat shows an increased amount of fat. An oral D-xylose absorption test reveals decreased reabsorption of xylose into the blood. Which of the following tests would be most useful in identifying the cause of the diarrhea?

1. Antigliadin antibodies ✔
2. Antinuclear antibodies
3. Fecal smear for leukocytes
4. Stool for ova and parasites
5. Stool osmotic gap

INCORRECT ✖

The correct answer is 1.

The patient has celiac disease, an autoimmune disease with antibodies directed against the gliadin fraction in gluten, which is present in wheat products. The antibodies cause an inflammatory reaction in the villi, resulting in villous atrophy (flat mucosa) leading to malabsorption of fat (greasy stools), carbohydrates, and protein. In addition, this patient has an increased quantitative stool test result for fat and abnormal results in the D-xylose reabsorption test, which is an excellent screening test for documenting small bowel disease as a cause of malabsorption. The vesicular lesion on the patient's elbow is dermatitis

herpetiformis, which is an autoimmune skin disease that has an almost 100% correlation with underlying celiac disease. Other antibodies present in celiac disease include anti-endomysial and anti-reticulin antibodies. The treatment of choice is to eliminate gluten from the diet.
(Choice 2) directed against nuclear proteins are not present in celiac disease.
(Choice 3) is used for evaluating diarrhea that may be caused by invasive microbial pathogens (e.g., Campylobacter jejuni, Shigella sonnei). The presence of leukocytes presumes an invasive enterocolitis and would not be expected in celiac disease.
(Choice 4) Testing the stool for ova and parasites is always recommended in the workup of a patient with chronic diarrhea. Giardiasis is the most common cause of chronic diarrhea associated with malabsorption; however, the presence of dermatitis herpetiformis excludes a parasitic cause of the chronic diarrhea.
(Choice 5) A stool sample to calculate the osmotic gap is used for high-volume diarrheal states when a secretory or osmotic type of diarrhea is suspected rather than malabsorption. The stool osmotic gap is obtained by measuring potassium and sodium in the diarrheal fluid, adding them together, and multiplying the number by 2 (i.e., 2 X {[potassium] + [sodium]}). This value is then subtracted from 300 mOsm/kg, which represents the osmolality of plasma. Secretory diarrheas are characterized by isotonic diarrheal fluid; therefore, the osmotic gap in stool will be less than 50 mOsm/kg. Causes of a secretory type of diarrhea include certain types of laxatives and enterotoxigenic bacteria such as enterotoxigenic Escherichia coli and Vibrio cholerae. Osmotic diarrhea is characterized by a hypotonic stool due to the presence of osmotically active solutes drawing more water than electrolytes out of the enterocytes. The classic example of an osmotic diarrhea is lactase deficiency, leading to an increase in lactose, which is osmotically active. The osmotic gap in osmotic diarrheas exceeds 100 mOsm/kg.

36. Question **1 points**

A 65-year old man presents to a primary care physician for his pre-Medicare physical examination. He is 5 feet, 10 inches tall and weighs 225 pounds, making his body mass index (BMI) 32.3; consequently, he is classified as being overweight but not obese. His physical appearance shows that he has a belly. In addition, the following laboratory data were obtained: fasting blood glucose value 115 mg/dL; blood pressure 180/95 mm Hg; total cholesterol 275 mg/dL, low-density lipoprotein 155 mg/dL, and high-density lipoprotein 25 mg/dL. The examining physician concluded that this man has metabolic syndrome and is likely to be pre-diabetic. Consequently, he subjected his patient to an oral glucose tolerance test by measuring the patient's fasting serum level, giving him a standard glucose drink, and then determining his blood glucose 2 hours later; the 2-hour value was 175 mg/dL, resulting in a diagnosis of prediabetes. He informed his patient that clinical studies have shown that the best way to reverse such a pre-diabetic state is by choosing to undergo a lifestyle change that includes eating a carefully controlled diet and exercising regularly; however, since many patients quickly fall away from such a régime, he also prescribed a drug that had been shown help prevent progression to full-blown diabetes when given to pre-diabetic patients. Moreover, this drug will not cause hypoglycemia, promote weight gain, or increase the risk of heart failure. Which one of the following drugs did the physician choose?

1. Sitagliptin phosphate
2. Glyburide
3. Metformin ✔
4. Rosiglitazone
5. Nateglinide

INCORRECT ✖

The correct answer is 3.

As indicated in the case study, this man shows the classic symptoms of metabolic syndrome (once called syndrome X). This includes a fasting glucose value higher than normal but below that required to diagnose diabetes (i.e., between 100 and 126 mg/dL), overweight, hypertension, and an abnormal serum lipid panel. Individuals having this syndrome feel well but are at high risk of developing full-blown diabetes. Even before becoming diabetic, they may be undergoing pathological changes that will lead to the typical complication of diabetes including retinopathy, nephropathy, and cardiovascular disease. Indeed, prediabetes likely should be considered a less severe form of diabetes rather than a unique condition in itself. The biguanide metformin is one of several oral diabetic drugs demonstrated to have the potential of reversing this pre-diabetic state; moreover, metformin operates primarily by inhibiting liver gluconeogenesis, as well as having a secondary ability to promote the uptake of glucose into muscle cells. In addition, it does not cause hypoglycemia or weight gain or increase the risk of heart failure, although it very rarely causes lactic acidosis. The potential ability of metformin to reverse the pre-diabetic state was evaluated in the Diabetic Prevention Program (DPP), a major clinical research program involving 27 research centers; in this study, overweight pre-diabetics persons were divided into three groups: the first group, called the lifestyle intervention group, received counseling, plus intensive training and supervised dietary and physical conditioning training as well as motivational therapy. The second and third group also received lifestyle counseling but not intensive training, supervision, or motivational therapy. The second group also received 850 mg metformin twice a day and the third group a placebo. In comparison to the placebo group, diabetes development was reduced 58% in the lifestyle intervention group and 31% in the metformin group.

(Choice 1) is the first drug in a relatively new drug class called dipeptidyl peptidase IV (DPP-4) inhibitors. DPP-4 normally breaks down the proteins that stimulate insulin production after a meal. Thus, a DPP-4 inhibitor permits insulin production to continue for a longer time, limiting the post meal increase in glucose. Sitagliptin phosphate does not cause hypoglycemia or weight gain; however, it has not yet been evaluated for its ability to reverse prediabetes, and sitagliptin may inflame the pancreas, conceivably worsening diabetes.

(Choice 2) is one of the many sulfonylureas that act by stimulating the pancreatic β cells to excrete more insulin. This class of drugs is liable to cause weight gain as well as bouts of hypoglycemia.

(Choice 4) and the other thiazolidinedione, pioglitazone, work by promoting the uptake of insulin by muscle cells, thereby decreasing insulin resistance in type 2 diabetics. It was reported at the 2008 meeting of the American Diabetic Association that pioglitazone lowered the progression from preto full diabetes by 81%; however, these thiazolidinediones promote water retention with concomitant weight gain and are suspected of inducing heart failure and therefore represent a false choice.
(Choice 5) and repaglinide are the two medications presently approved for treatment of type 2 diabetes. These drugs, like the sulfonylureas, stimulate the β cells to produce insulin; in contrast to the sulfonylureas, however, they only do so when blood glucose concentrations are elevated. Thus, they are taken usually three times a day, before a meal; as a result, they reduce the postprandial glucose high. Like sitagliptin phosphate, neither nateglinide nor repaglinide have yet been evaluated with respect to their potential for reversing prediabetes.

37. Question **1 points**

A bereaved family just returned from a funeral for their 20-year-old son. In her grief, the mother began to reminisce to her mother. “He was such a healthy baby until he reached his third birthday. Then he stopped running and started walking like a duck. Next, he couldn’t stand without climbing up on somebody or something, and by his 11th birthday, we had to get him a motorized wheelchair. Even then he was doing well, finished high school, enrolled in City College, was a member of the chess team, and took an interest in politics. Then he caught what seemed just to be a cold and died.” The grandmother replied, “Yes, it is so tragic. I can’t understand what happened. Nobody else in the family ever had anything like his disease. Not his sisters, your sisters, or my sisters.” Which of the following diseases did this young man have?

1. Becker muscular dystrophy
2. Myotonic muscular dystrophy
3. Facioscapulohumeral muscular dystrophy
4. Limb girdle muscular dystrophy
5. Welander distal myopathy
6. Duchenne muscular dystrophy ✔

INCORRECT ✖

The correct answer is 6.

At least nine major variant types of muscular dystrophy exist, with several subtypes. The common feature is a progressive form of muscular weakness. The most common of these is Duchenne, the disease that killed the boy described in the vignette. It has an estimated incidence of 1 case per every 3,500 male live births and is due to a major deletion on the

short arm of the X-chromosome (Xp21). It is inherited as a sex-linked recessive trait and results in the nonproduction of dystrophin, a very large protein (427 kDa) that is part of the contractile apparatus of muscle cells. In addition to making the muscles weak, the absence of a dystrophin molecule makes the muscle cell leaky, which leads to a marked increase in serum creatine kinase activity and contributes to the ultimate death of muscle cells. Affected boys seem normal until the age of 2–5 years, when they develop a peculiar duck-like waddling gait caused by weakness of the pelvic girdle muscles. The weakness then progresses throughout the legs, forcing them into a wheelchair by their teens, and then to the upper body. During the early period, their lower legs often increase in diameter, not from muscle, but from fat and connective tissue causing pseudo-hypertrophy. By their late teens, the respiratory muscles and/or heart also weaken, and these patients succumb to a respiratory disease or cardiac failure not long afterward. There is no treatment. The literature often reports mild retardation (an average IQ of 85), but there are many exceptions to this, as in the chess-playing boy described here.

(Choice 1) also results from a mutation in the dystrophin gene, but the mutation permits production of a defective dystrophin protein that has limited function. The result is a muscular dystrophy similar to Duchenne but not as severe. First symptoms usually occur during the late teens or early 20s, and affected persons are able to walk into their 30s. However, they too will eventually die from either respiratory or cardiac failure. Duchenne muscular dystrophy is also inherited as an X-linked recessive disorder.

(Choice 2) The incidence is about 1 case per 30,000 live male births. Type 1 myotonic muscular dystrophy is the most common adult dystrophy and takes its name from the tendency for affected persons have progressive muscle wasting and weakness, coupled with myotonia, the inability to relax muscles after use; thus, for example, a person may not be able to put down a fork after using it. At least two distinct forms of myotonic muscular dystrophy exist, type 1 (DM1) and type 2 (DM2); DM1 accounts for as many as 98% of the cases and affects about 1 in 8,000 individuals. Both forms are inherited as autosomal dominant diseases and DM1 in particular shows anticipation the disease appears earlier and become more severe with each successive generation. DM1 is caused by a trinucleotide repeat on chromosome 19 that result in a defective myotonin protein kinase, whereas DM2 is caused by a tetra-nucleotide repeat on chromosome 3. Additional poorly characterized variants of myotonic muscular dystrophy might also exist. Diagnosis of the myotonic muscular dystrophies is difficult to make because not only do symptoms vary from family to family, the range and severity of symptoms can vary greatly among patients even in the same family. At one extreme, a patient may only develop mild muscle weakness and/or cataracts late in life, whereas at the other extreme, a child might be born with the congenital form of the disease and have early-onset life-threatening disease. All patients with the condition are at risk of severe reactions to anesthesia and should be monitored carefully if subjected to anesthesia.

(Choice 3) is an autosomal dominant condition that usually starts in the teens or early 20s. It first affects the face and shoulder girdle and then the pelvic girdle, legs, and abdomen. Clinically, its effects vary from very mild to severe; about 50% of patients can walk until they die at a normal age.

(Choice 4) can be inherited in an autosomal dominant or more commonly in an autosomal recessive fashion. First symptoms usually start in the teens or early adulthood as weakness in the hips. It then slowly progresses, first to the shoulders, then to the arms and legs. Some

20 years after the first onset of symptoms, walking becomes very difficult.
(Choice 5) is a rare autosomal dominant condition most commonly is found among persons of Swedish or Finnish ancestry, having a prevalence of about 1 in 4,000 in mid-Sweden. It is one of a group of heterogeneous myopathies classified as distal myopathies, characterized clinically by muscular weakness and atrophy beginning in the hands and feet. The disease generally then progresses in severity and to muscles elsewhere in the body. As many as two dozen suspected distinct types of distal myopathies have been described (at least five of which have been identified as mutations in a specific gene), making diagnosis difficult. Most have adult onset, a major exception being Laing myopathy, which can be identified in infancy. In addition, individuals in the same family can be affected to markedly different degrees. One member may live a long, essentially normal life with little more than what appears to be arthritis of the hands, whereas another may be bedridden by mid-adulthood.

38. Question **1 points**

A 74-year-old widow who lived alone fell at home and hurt her left wrist. She could immediately see that it was broken because of the way it dangled. Consequently, she called 911. The paramedics set her arm in splints and took her to the nearest emergency clinic. While the attending physician treated the fracture, he also took a history. Among the questions he asked was: “Why do you think you fell?” She responded that she became dizzy; she felt the room was spinning while she was standing in one place; consequently, she lost her balance. The physician probed further and subsequently discovered that she had been having similar spells of dizziness intermittently at various times both during the day or night for the past several months. The dizziness could occur while she was lying down, sitting, or standing without her changing her position. He further determined that her left ear often felt full and that she sometimes had a faint but annoying ringing in her ears. She denied ever vomiting during these dizzy spells but did say she often felt nauseated. Her dizziness was most likely caused by which of the following?

1. Shy-Drager syndrome
2. Ménière disease ✔
3. Essential hypertension
4. Orthostatic hypotension
5. Benign positional vertigo

INCORRECT ✖
The correct answer is 2.
Ménière disease is one of a host of conditions that can cause dizziness. Characteristics in the case history described that point to Ménière disease are the description of the room

spinning while the patient stood still, tinnitus, and a sensation of fullness in one or both ears. It is also often also accompanied by nausea and vomiting. Since there is no specific test, it is diagnosed by clinical symptoms and response to treatment. Specifically, acute attacks usually respond to the antivertigo drug meclizine and/or benzodiazepines but not to treatment used to prevent other causes of dizziness.

(Choice 1) Shy-Drager syndrome; aka, neurologic orthostatic hypotension or multiple system atrophy) is a rare disease that, among a host of other symptoms, causes a feeling of dizziness. The disease starts gradually, often manifesting with frequent falls caused by dizziness due to a drop in blood pressure upon standing up. It progresses into a parkinsonian-like condition that includes slowness of movement, muscle rigidity, bladder dysfunction, and poor balance; ultimately, it causes death. Commonly, it affects males more than females, and symptoms start at about the age of 50–60 years. The patient described is female, and by the age of 74 years, had she had Shy-Drager syndrome she would likely had symptoms more severe than dizziness.

(Choice 3) should not of itself cause dizziness, nor does the case history mention hypertension.

(Choice 4) is a common cause of dizziness provoked by rapidly standing, particularly from a supine position; typically, the patient feels faint. Moreover, the dizziness described by the lady in this question occurred while she was lying down, sitting, or standing, and was not provoked by a change in position.

(Choice 5) is the most common cause of peripheral vertigo. Normally, tiny mineral particles help maintain balance by rotating within the semicircular canals. In benign positional vertigo, some of these get stuck in one place. Most patients get relief by body repositioning maneuvers that can be performed in the physician's office or even at home. Benign positional vertigo is not associated with tinnitus (although a patient may have had tinnitus before being troubled by vertigo), or with a feeling of fullness in an ear; nor is it likely a patient will describe the room as spinning. A patient with benign positional vertigo is more likely to describe the dizziness as an unsteady feeling and/or faintness rather than a spinning room.

39. Question **1 points**

A 40-year-old man came to see his primary care doctor as he had been feeling very fatigued of late. He stated that he was tired all the time, had difficulty walking the distance that he could only a month or so ago, and he also noticed some swelling of his feet, usually at the end of the day. Physical examination revealed a medium-built well-nourished man with conjunctival pallor. The sclera appeared mildly icteric. His temperature was 98.6°F (37.0°C), pulse 88/min regular, respirations 18/min, and his blood pressure was 120/86 mm Hg. Oxygen saturation using room air was 90%. His lungs were clear to auscultation, bilaterally; he had a soft murmur in the precordial area without a thrill, and no gallops or rubs were noted. The abdomen was soft to palpation, and he had mild tenderness without guarding or rigidity in the right upper quadrant. No organomegaly was noted, and bowel sounds were present in all four quadrants. A complete blood count revealed a low hematocrit and a corrected reticulocyte count of greater than 3%. Which one of the following changes in serum levels also would most likely occur?

1. Conjugated bilirubin, haptoglobin, and lactic acid dehydrogenase levels would all be elevated

2. Conjugated bilirubin would be elevated; haptoglobin and lactic acid dehydrogenase levels would be decreased

3. Unconjugated bilirubin and haptoglobin levels would decrease; the lactic acid dehydrogenase level would increase

4. Unconjugated bilirubin, haptoglobin, and lactic acid dehydrogenase levels would all increase ✔

5. Only the haptoglobin level would increase

INCORRECT ✖

The correct answer is 4.

This patient has a hemolytic anemia. Hemolytic anemia occurs when more red blood cells are destroyed than are being produced. It could result from immune or nonimmune disorders; the defect could be intrinsic or extrinsic. Causes include intrinsic red cell anomalies; premature destruction of otherwise normal cells, as occurs in mismatched transfusion; drug interaction; infection due to some strains of streptococci; and hypersplenism. Intrinsic abnormalities of red blood cells that may cause a hemolytic anemia include hereditary spherocytosis, in which there usually is an abnormality in spectrin, a protein that provides most of the scaffolding for the red cell membrane and enables it to deform; deficiencies in one of several of the enzymes involved in glucose metabolism; and abnormal hemoglobins, such as hemoglobin S, which causes sickle cell disease, an autosomal recessive disorder leading to sickling of red cells under conditions of local hypo-oxygenation. A peripheral blood smear will help distinguish between spherocytes and sickle cells. If sickle cells are found, hemoglobin electrophoresis is required to establish whether the sickling is due to sickle cell disease. In hemolytic anemia, the levels of unconjugated bilirubin, haptoglobin, and lactic dehydrogenase are increased. Bilirubin is a breakdown product of hemoglobin and is usually transported to the liver via the circulation. For this to happen, bilirubin has to be noncovalently associated with albumin. This complexed form is called unconjugated or indirect bilirubin. Once it reaches the liver, it is metabolized and albumin is no longer required. Most of this bilirubin freed from albumin is covalently bound to glucuronic acid is now called conjugated or direct bilirubin. However, in cases of hemolysis, a large quantity of unconjugated bilirubin remains circulating in the blood because the liver is unable to conjugate such a large quantity; hence, the level of circulating unconjugated bilirubin increases. Haptoglobin is a circulating protein and is an acute-phase reactant, meaning its concentration is increased in the serum in conditions in which stress, infection, or inflammation occurs. In cases of hemolysis, haptoglobins bind the hemoglobin that is released, thus minimizing its accumulation in the plasma. The hemoglobin–haptoglobin complex is then eliminated by the reticuloendothelial system. Lactic dehydrogenase is present in the cytoplasm of red cells and is released into the circulation when cells undergo hemolysis, resulting in its elevation.

(Choices 1,2,3 & 5) All are incorrect.

40. Question

1 points

A 77-year-old man who prides himself as having been in prime physical health all his life despite seldom seeing a doctor, and who claims the only medication he ever takes is aspirin, suddenly is plagued by severe headaches, vertigo, and a loss of balance that makes standing and walking difficult. Consequently, he has a fall and bumps his head. His worried daughter takes him to the nearest emergency department, where he is examined. He is found to be a thin man who is relatively muscular for his age. Although still unable to stand or walk without losing his balance, there is no evidence of muscle weakness. He remains fully conscious, is fully cognizant of his surroundings, and can answer questions quickly and lucidly. His heart and lungs are normal, as is an electrocardiogram (ECG). A non fasting finger stick blood glucose determination reads 152 mg/dL of glucose. His blood pressure remains between 165 and 185 mm Hg systolic over 75–85 mm Hg diastolic. Which of the following is the most likely underlying cause of his problem?

1. An epidural hemorrhage
2. Ménière syndrome
3. Isolated systolic hypertension ✔
4. A cerebral infarct
5. A pulmonary thromboembolism

INCORRECT ✖

The correct answer is 3.

Isolated systolic hypertension is a condition that often affects the elderly; in fact, it is so common that it was once believed that normal systolic pressure is equal to 100 plus a person's age. However, trial studies have conclusively demonstrated that elevated systolic blood pressure is a more significant underlying cause of cardiac disease and stroke than is elevated diastolic pressure; 30% of women and 20% of men older than 65 years have this condition. The underlying reason is a loss of arterial elasticity that occurs with aging, and the condition becomes more common with advancing age. The man described most likely is suffering from a cerebellar stroke, which typically is associated with headache and ataxia. **(Choice 1)** is a consequence of trauma and could conceivably have resulted from his fall. However, his fall occurred after the symptoms first appeared; typically, an epidural hematoma is characterized by coma after a lucid interval, and there is no mention of loss of consciousness.

(Choice 2) is a cause of vertigo induced by distention of the endolymphatic compartment of the inner ear. Although usually idiopathic, it can be caused by head trauma or syphilis. It is not age associated, and there is no evidence presented in the vignette that this patient might have Ménière syndrome.
(Choice 4) would not have only primary symptoms relating to ataxia but would rather have loss of muscular strength or cognitive function.
(Choice 5) is usually accompanied with signs of dyspnea, chest pain, hemoptysis, and/or syncope. Except for syncope, the man had none of these symptoms. Moreover, there is no indication of a predisposition for venous thrombosis, and the one drug this man uses is aspirin.

41. Question **1 points**

A 25-year-old man presents to the emergency room with a history of swelling around the left eye, which has become very painful. He states that initially he had noted double vision and that it hurts to move his eye. He was in considerable distress due to pain. The vital signs were as follows: temperature 101°F (38.3°C), pulse 86/min regular, respirations 18/min, blood pressure 128/78 mm Hg and, oxygen saturation 98% on room air. There was considerable swelling around the left eye, which was boggy and tender to touch. Ocular movements were present in all directions but were painful. The pupils were equal and reacting appropriately to light and accommodation, and funduscopy was unremarkable. The most likely cause for this problem is:

1. Cavernous sinus thrombosis
2. Corneal ulcer
3. Sinusitis ✔
4. Orbital abscess
5. Ocular trauma

INCORRECT ✖

The correct answer is 3.

This patient has orbital cellulitis, which is most commonly due to sinusitis. Patients will often give a history of prior upper respiratory infection. Orbital cellulitis can lead to life-threatening complications, including cavernous sinus thrombosis and orbital abscess. This is an emergency and should be treated with intravenous broad-spectrum antibiotics. Sinusitis, if present, should be treated,
(Choice 1) Although cavernous sinus thrombosis is a potential complication of orbital cellulitis, this patient as not yet developed this condition. In addition to fever and diplopia, it usually presents with a history of nausea, vomiting, and headache. Alteration in mental

status also may be present, and the pupil is dilated.
(Choice 2) is associated with blepharospasm (the eyelid is squeezed shut, as if squinting), epiphora (increased tear secretion), and ocular pain. Diplopia and fever are not features.
(Choice 4) results from ulitis (inflammation of the gums) and is a potential complication of orbital cellulitis.
(Choice 5) The patient does not give a history of trauma, and if trauma were responsible for the symptoms, ocular hemorrhage would probably have been associated with it and noted on examination.

42. Question **1 points**

An 82-year-old retired university professor of Scott-Irish ancestry has had gastroesophageal reflux disease (GERD) and hypertension for a number of decades. Lately, he has been having increasing difficulty walking; his gait has become more and more shuffling and hesitant. More recently, he has also complained of excessive fatigue; for example, he might doze off in his chair while eating breakfast. His wife thought this might be because he did not get a good night's sleep in fact, she complained that she couldn't sleep well either because of his loud snoring. His primary care physician is treating his GERD with omeprazole and is using six different antihypertension drugs (enalapril, labetalol, clonidine, doxazosin mesylate, amlodipine besylate, and hydrochlorothiazide) to control his systolic hypertension, but his systolic pressure remains, at best, in the 140–160 mm Hg range. Frustrated, the physician arranges for his patient to be tested for possible obstructive sleep apnea, which theoretically could cause fatigue and muscle weakness, and contribute to the hypertension. Testing shows that he had an inadequate amount of rapid eye movement (REM) sleep, and his oxygen saturation dropped below 90% on a regular basis. He was provided with a continuous positive airway pressure (CPAP) device, so that he could treat himself at home. However, his systolic blood pressure stubbornly remained above 140 mm Hg, and he continued to walk poorly and doze off during the day.

Looking for some other source for his patient's hypertension and fatigue, the physician ordered a battery of blood tests including a complete blood count (CBC). Which of the following patterns could best explain this patient's symptoms?

1. Normal hemoglobin levels and a mixture of normal-looking red cells mixed in with some abnormally large erythrocytes, some poikilocytes, a few hypersegmented neutrophils, and a reduced the reticulocyte count ✔

2. Normal red cells mixed in with a substantial fraction of schistocytes and normal leucocytes and a decrease reticulocyte count

3. Primarily spherocytic red cells, normal white cells, and a decreased reticulocyte count

4. Normal red cells mixed in with some sickled cells and a decreased reticulocyte count

5. A reduced number of normal red cells with normal white cells and a decreased

reticulocyte count

INCORRECT ✖

The correct answer is 1.

The pattern described in answer A describes a macrocytic/normochromic anemia (i.e., an anemia that produces abnormally large red cells that have normal hemoglobin content); this is also known as a megaloblastic anemia. This condition occurs whenever DNA synthesis is inhibited in an otherwise viable developing cell. The inhibition of DNA synthesis delays nuclear maturation and induces an imbalanced rate of development in which cell division is inhibited, thus permitting the erythroblasts in the bone marrow to grow to an abnormal size and often an aberrant shape. (The generic term for such irregularly shaped cells is poikilocytes.) Although cell division is inhibited cytoplasmic functions continue permitting the maturing red cell to take up hemoglobin. As development continues, the unusually small nucleus of the blast cell is lost, and the malformed red cell is transport into the circulation, where it is readily recognized. Folate is required for DNA synthesis, and consequently, this type of abnormal development always occurs under conditions of absolute or functional folate deficiency. Functional deficiency of folate can occur when sufficient vitamin B_{12} is not available because vitamin B_{12} is a cofactor in the homocysteine methyltransferase reaction, which methylates N_5-methyl tetrahydrofolate to form tetrahydrofolate, an active form of folate used in DNA synthesis. (N_5-methyl tetrahydrofolate is the end-product of all metabolically active forms of folate and has no other catalytic function; thus if vitamin B_{12} levels are insufficient to permit this methylation reaction, a functional deficiency of folate will occur, provided dietary intake of folate is not adequate enough to overcome its loss via conversion of the active forms to the N_5-methyl tetrahydrofolate derivative.)

As ingested, dietary vitamin B_{12} passes through the stomach, where it attaches to a glycoprotein called the intrinsic factor, which is synthesized by the gastric parietal cells; this binding requires a low pH. The vitamin B_{12}-intrinsic factor complex then binds to specific receptors in the mucosal cells of the ilium and subsequently is transported into the general circulation. Pernicious anemia results when this complex mechanism of vitamin B_{12} absorption goes awry. Such malfunction may result from an autoimmune reaction disabling the production or function of the intrinsic factor, an autoimmune reaction or surgery causing the loss of parietal cells, or a lower than normal gastric pH. Pernicious anemia has been reported to be most commonly found among persons of Celtic or Scandinavian ancestry but in the past few decades, people from other ethnic groups have been reported to have a similar prevalence. This may be because in modern developed societies the hematologic symptoms of pernicious anemia have become more difficult to observe because they are commonly made imperceptible by excess folate in the diet, which makes the conversion to N_5-methyl tetrahydrofolate largely inconsequential. One might think this would make the reduced ability to absorb vitamin B_{12} unimportant; however, this vitamin has one additional function in normal metabolism and that is as a cofactor in the methylmalonyl CoA mutase reaction that converts methylmalonyl CoA to succinyl CoA. This reaction permits the conversion of propionyl CoA to succinate; this propionyl CoA is produced by the final β-oxidation step during the oxidation of odd-carbon fatty acids, as well as from valine, leucine, and methionine. Thus, a deficiency of vitamin B_{12} inhibits the methylmalonyl CoA mutase

reaction and leads to an accumulation of propionyl CoA, which can get incorporated into fatty acids during fatty acid synthesis; this will produce odd-carbon fatty acids that then get incorporated into neural myelin causing neuropathy, a relatively early symptom of which is loss of proprioceptor function. However, over the long-term it may cause more severe symptoms, including psychiatric symptoms resulting in senile dementia. In addition to showing some hematologic symptoms, namely signs of a macrocytic-normochromic anemia, the patient described shows signs of neuropathy, namely loss of proprioceptor function marked by a shuffling hesitant gait. In addition, he might be genetically prone to develop pernicious anemia because of his Celtic ancestry and because his gastroesophageal reflux disease (GERD) is being treated with a protein pump inhibitor.
(Choices 2,3,4 & 5) are incorrect because the root part of the answer is irrelevant and because each condition described should result in increased not decreased reticulocyte formation.

43. Question **1 points**

A 43-year-old Caucasian man of northern European descent had been feeling a rapidly escalating degree of fatigue for the past several weeks and consequently consulted his primary care physician. Upon taking a history, his physician confirmed that, in addition to feeling tired, the patient has noted a shortness of breath and heart palpitations. Laboratory analyses showed low serum hemoglobin and haptoglobin levels and raised serum reticulocyte, lactic acid dehydrogenase, and bilirubin concentrations. Deducing that her patient may have a hemolytic anemia, the physician referred him to a hematologist, who determined that there was no family history of anemia. The hematologist asked the patient to void a midday urine sample into a prelabeled Erlenmeyer flask, and to also provide her with a second urine sample obtained immediately after rising in the morning; she also and told him to bring both samples into her office the following day. The patient did as requested; the midday urine sample had a deep orange-yellow color typical of concentrated normal urine, but the early morning sample was a bright reddish color. The patient most likely suffered from which one of the following conditions?

1. A drug-induced non autoimmune hemolytic anemia
2. Paroxysmal nocturnal hemolytic anemia ✔
3. Paroxysmal cold hemoglobinuria
4. Warm autoimmune hemolytic anemia
5. Acute intermittent porphyria

INCORRECT ✖

The correct answer is 2.

Paroxysmal nocturnal hemolytic anemia (PNHA) is a rare acquired form of hemolytic anemia. It is characterized by hemolysis that typically occurs during the night, resulting in excretion of early morning urine colored dark red by hemoglobin released from lysed red cells. The first symptoms may present at any age from infancy into the 80s, but it most frequently affects adults between 17 and 75 years old, with a mean age of presentation of 42 years. It is caused by the loss of membrane surface proteins due to an apparent mutation that inhibits the ability to synthesize glycosyl-phosphatidylinositol, which normally serves to anchor these surface proteins to the cell. The responsible gene is called phosphatidylinositol glycan class A (PIGA) and the mutations may be of several types, including frame shifts, point mutations, and deletions. This defect is believed to be a stem cell mutation because all cells in the hemopoietic line are affected; it is hypothesized that the mutations are caused by autoimmune reactions. The critical membrane proteins involved in PNHA symptomlogy normally react with members of the complement family, including those that normally act to impede the amplification of complement action; consequently, such a mutation acts to inhibit the normal dampening control in the complement cascade, permitting the action of complement to run amok and resulting in cell lysis. Because all hematopoietic cell lines are affected, symptoms extend beyond lysis of erythrocytes, causing a generalized pancytopenia that reduces the number of white cells and platelets, as well as causing an anemia. Thus, in addition to the anemia, thrombi often occur, particularly in large veins; these may cause severe disease, resulting in death. In theory, the ideal treatment is stem cell replacement; unfortunately, this process is not yet practical. Presently, treatment includes anticoagulants plus an anticomplement antibody called eculizumab. Use of this drug dramatically improves all almost all symptoms of PNHA.

Whenever anemia is found in a patient, it is important to ascertain whether the anemia is due to a disorder in the bone marrow, to hemolysis, or to loss of blood. A corrected reticulocyte count will help make the differentiation. The formula for a corrected reticulocyte count is the patient's reticulocyte count, multiplied by the patient's hematocrit, divided by the expected hematocrit. If the corrected reticulocyte count is below 2%, the most likely cause of anemia is decreased red blood cell production due to a hypoproliferative bone marrow. On the other hand, if the corrected reticulocyte count is greater than 3%, the most likely cause is hemolysis. In this patient, who has hemolytic anemia, it is important to establish if the etiology is due to an immune or a nonimmune disorder. This can be achieved by the Coombs test.

(Choice 1) The classic example of a drug-induced non autoimmune hemolytic anemia is a glucose-6 phosphate dehydrogenase deficiency. It can be ruled out in the case described because the onset of glucose-6 phosphate dehydrogenase deficiency is sudden and dependent upon ingestion of a drug, rather than a gradually worsening fatigue; moreover, the patient most likely would be African American or of Mediterranean descent.

(Choice 3) is a rare condition characterized by an abrupt onset of a severe hemolytic anemia and hemoglobinuria when a patient is exposed to cold temperatures. (The hemolysis usually occurs in a limb or digit exposed to a cold temperature.) Induction of this phenomenon involves the attachment of a foreign antibody to the red cell, which is promoted by the reduced temperature; hemolysis is then induced by activation of the complement system when cells are warmed. The phenomenon commonly manifests itself as a sudden

transitory development induced by infections, usually a postviral episode. Before the advent of antibiotics, it was more commonly observed, often among patients suffering the tertiary stage of syphilis.
(Choice 4) is the most common type of hemolytic anemia and is usually found in mature adults. In this condition, the patient's body makes auto-antibodies against their own red blood cells, and hemolysis occurs under warm conditions (i.e., normal body temperature). Diagnosis is generally made by the direct Coombs test.
(Choice 5) is a rare dominant disease caused by a deficiency of porphobilinogen deaminase. Insufficient activity of this enzyme leads to the accumulation of porphobilinogen in the cytoplasms of cells in various organs in the body. This accumulation may be influenced by various hormones and external stimuli, causing intermittent attacks. These attacks may include various digestive, muscular, or even mental aberrations and the voiding of port wine–colored urine, which turns to purple when exposed to ultraviolet light for a period of time.

44. Question **1 points**

A 35-year-old Caucasian man with neurofibromatosis presents to his physician with a complaint of nausea and vomiting, vertigo, nystagmus, tinnitus, and nerve deafness in the right ear. These symptoms had gradually worsened over the past month or so. Upon examination, the physician noted hemianesthesia on the right side of the face. Which of the following is the most likely diagnosis?

1. Mastoiditis
2. Cerebellar tumor
3. Vertebrobasilar arterial insufficiency
4. Glioblastoma multiforme
5. Acoustic Schwannoma ✔

INCORRECT ✖

The correct answer is 5.

An acoustic schwannoma is associated with tinnitus, nausea and vomiting, vertigo, nystagmus, and eighth-nerve deafness. It arises in the cerebellopontine angle and may involve the trigeminal nerve, producing ipsilateral sensory changes in the face. It is most commonly a schwannoma a benign which is a benign, encapsulated tumor arising from Schwann cells. Patients with neurofibromatosis have an increased incidence of acoustic schwannomas.
(Choice 1) Untreated mastoiditis that erodes through bone to produce acoustic nerve damage is rare.

(Choices 2 & 3) (Choice 2) and vertebrobasilar artery insufficiency **(Choice 3)** produce ataxia. Furthermore, they are not usually associated with eighth-nerve damage.
(Choice 4) is the most common primary malignancy of the brain in adults and usually involves the frontal lobes. It would not be expected to produce eighth-nerve damage.

45. Question **1 points**

A 45-year-old woman sees her primary care doctor for fatigue. She states that she used to be able to walk a mile “with ease,” but of late has been having problems doing so. She feels tired and winded, and has noticed some problems eating as her tongue “feels raw.” This patient most likely has which one of the following conditions?

1. Microcytic anemia
2. Macrocytic anemia ✔
3. Thalassemia
4. Sideroblastic anemia
5. Anemia of chronic disease

INCORRECT ✖

The correct answer is 2.

This patient has pernicious anemia due to vitamin B_{12} deficiency. In younger patients such as this, the most common cause of pernicious anemia is an autoimmune reaction in which antibodies destroy the stomach’s parietal cells, which normally manufacture intrinsic factor. Intrinsic factor is required for B12 absorption in the terminal ileum. As in this case, patients may have glossitis, as well as neurological symptoms such as tingling in the fingers, loss of vibration and position sense, and in extreme cases, dementia. Other causes of B12 deficiency anemia in include inadequate dietary intake of vitamin B_{12}, alcoholism, partial gastrectomy, malabsorption syndromes, Crohn’s disease, and infestation with the fish tape worm (Diphyllobothrium latum), which is present in fresh water fish in the northern hemisphere. Antiparietal cell antibodies will be present in autoimmune cases of pernicious anemia. In the presence of macrocytic anemia, it is important to check both B12 and folate levels, as folic acid deficiency could lead to macrocytic anemia as well. Red blood cell folate level is more accurate than serum folate level. Whether a macrocytic anemia is due to lack of B12 or folic acid can be resolved by ascertaining the levels of serum methylmalonic acid and homocysteine. Both levels are increased in B12 deficiency, but in the case of folic acid deficiency, only homocysteine levels are elevated. Methylmalonic acid is an intermediate in fatty acid metabolism that is derived from malonate, while homocysteine, an amino acid not found in diet, is derived from methionine. It is involved in the metabolism of cysteine. A

Schilling test will distinguish between nutritional or absorptive cause of macrocytic anemia. Conversely, it is also possible that high intake of folate will prevent signs of a macrocytic anemia even if a patient has a vitamin B_{12} deficiency. Thus, one should pay careful attention to neurological symptoms that may be the only sign of a vitamin B_{12} deficiency and could easily be overlooked; unfortunately, these may in the long run cause severe permanent neurological damage, including psychological aberrations.

(Choice 1) is seen in iron deficiency that can result from inadequate intake or absorption of iron, or excessive blood loss, with the most common cause being inadequate intake. Clinical features include cheilosis, in which the lips are cracked at the angles of the mouth, dysphagia, brittle nails, and in extreme cases, koilonychia, also known as spooning of the nails.

(Choice 3) is the most common genetic disorder worldwide. It is due to abnormal hemoglobin. Mature hemoglobin is a tetramer of two and two α chains. If there is an aberration in this combination, interaction with oxygen will result in precipitation of hemoglobin and destruction of the abnormal red blood cell by the spleen, with resultant hemolytic anemia. Patients will have microcytic hypochromic red blood cells in the peripheral smear. Thalassemia can affect either the α or β chains, hence hemoglobin electrophoresis is required to uncover the malady.

(Choice 4) is usually an acquired form of anemia in which there is an inability to incorporate heme into protoporphyrin to create hemoglobin. Causes include myelodysplasia, which may progress to acute leukemia; chronic alcoholism; and lead poisoning. Patients have mild to moderate anemia, and usually present with fatigue. Mean corpuscular volume (MCV) is usually normal. In some instances, it may be decreased, causing confusion with iron deficiency anemia. The distinction can be made by the peripheral blood smear, which shows two kinds of red blood cell populations, some normal and others hypochromic. In addition, iron stores are increased due to the primary anomaly, which is that heme cannot be adequately utilized. The diagnosis is best made by examining the bone marrow, which will reveal ineffective erythropoiesis as noted by the presence of marked erythroid hyperplasia, increased iron stores, and the presence of ringed sideroblasts. The serum iron and transferrin saturation will be markedly elevated, and iron may be deposited in mitochondria as well. Sideroblastic anemia due to lead poisoning will reveal basophil stippling in the red blood cells and elevated lead levels in the serum.

(Choice 5), as the name suggests, occurs in chronic systemic disorders such as infection, inflammation, hepatic disease, and malignancy. Chronic renal failure is also a notable cause. In cases other than chronic renal failure, anemia results from reduced red cell survival rate, and the inability of the bone marrow to step up production as iron is stashed away in the reticuloendothelial system where it has been delivered by haptoglobin. In chronic renal failure, on the other hand, anemia is due to inadequate production of erythropoietin, which is essential to prime the bone marrow to produce red blood cells. The diagnosis of anemia of chronic disease is confirmed by the presence of low serum iron, low total iron binding capacity, and normal or elevated levels of ferritin, and in the case of anemia of chronic disease, low erythropoietin levels.

46. Question

1 points

A recently retired 67-year-old man makes an appointment to see his physician because his hands are beginning to tremble in an uncontrollable manner. During the examination, the physician notes that he does have an obvious resting tremor of both hands. However, he had no difficulty in picking up a quarter, and during that process the tremor ceased. When asked to hold his hand out, palm up, his fingers underwent an involuntary movement as if he was rolling a pill across his palm. He states that although the tremor is annoying and sometimes embarrassing, it makes little difference in his lifestyle; it has not even interfered with playing golf. At this time, which of the following is the best first step in treatment?

1. Amantadine
2. An anticholinergic drug
3. Levodopa
4. Carbidopa
5. A dopamine agonist
6. Selegiline
7. Watchful waiting ✔

INCORRECT ✖

The correct answer is 7.

This man is suffering from early parkinsonism. Most commonly, this is due to Parkinson disease, a condition of unknown etiology that generally starts between the ages of 45 and 65. Parkinson disease is a heterogenous and progressive condition that has tremor, rigidity, bradykinesia, and postural instability as its cardinal features. Cases tend to fall into one of two major subtypes. In one group, tremor is the major symptom; in the other, postural instability and gait difficulty predominate. Even though symptoms do overlap, most patients clearly fall to the greatest extent into one or the other subcategory. Patients with predominantly tremor-type symptoms tend to have a slower progression of symptoms, have less trouble with bradykinesis, and less often develop severe mental symptoms. Other symptoms in more advanced cases include infrequent blinking, a blank stare, a shuffling gait with rapid acceleration and difficulty stopping once started, increased salivation, and severe depression or even dementia. No one patient needs to develop all symptoms to meet the diagnostic criteria, and patients who have early symptoms of gait disturbances progress more rapidly and are more apt to develop the more severe symptoms. The underlying pathology is dopamine depletion due to degeneration of the nigrostriatal system. This leads to an imbalance between acetylcholine and dopamine neurotransmission. The cornerstone of treatment is dopamine replacement coupled to blockage of the acetylcholine system. Unfortunately, resistance to ceratin key drugs develops with use. Therefore, an attempt is

made to refrain from using them as long as possible. Thus, watchful waiting is typically used until symptoms interfere with the patient's normal lifestyle. This patient clearly states that his tremor does not impinge upon his normal lifestyle.

(Choice 1) Once it is decided that some medical intervention will be of value, amantadine is typically the first drug to be used while symptoms are still minimal. This antiviral drug helps to improve muscle control and reduce stiffness; its mode of action is not understood, but it is hypothesized to help release dopamine from nerve endings. Its effect is always less than profound, and the benefit further decreases with use.

(Choice 2) Anticholinergics also are used to treat early parkinsonism, sometimes in conjunction with amantadine. Again, drug resistance develops, and these drugs are generally administered in increasing doses until the adverse effects outweigh the benefits. Thus, eventually, it becomes necessary to use the most effective treatment namely, dopamine replacement. However, dopamine cannot cross the blood brain barrier.

(Choices 3 & 4) Consequently, it is usually administered as Sinemet, a preparation that contains levodopa and carbidopa in fixed proportions. The levodopa is converted to dopamine in the body, and the carbidopa inhibits the enzyme that converts levodopa to dopamine. But carbidopa cannot cross the blood–brain barrier; thus, by administering the two together there is both more levodopa available, and some of the adverse peripheral effects of dopamine such as nausea, vomiting, hypotension, and cardiac irregularities are avoided. Long-term administration of levodopa induces untoward central nervous system effects including dyskinesias, restlessness, confusion, and behavioral changes. Still later during therapy, the so-called on–off phenomena may occur, in which severity of parkinsonism may quickly increase at any time of the day.

(Choice 5) Dopamine agonists that act directly on dopamine receptors were once only used to reduce the symptoms associated with this on–off action associated with long-term levodopa use. However, with the advent of newer dopamine agonists that are not derived from ergot, such agonists are now used to help treat early Parkinson disease and to keep the levodopa dose (via Sinemet) at a minimal level as long as possible.

(**(Choice 6)**) is an irreversible monoamine oxidase B (MAO-B) inhibitor. MAO-B selectively deaminates dopamine and phenethylamine (chocolate's amphetamine). Selegiline is sometimes used as an adjunct treatment, with the idea of reducing symptom fluctuations associated with long-term levodopa therapy. During the past decade, an ever-increasing number of otherwise refractory patients have been treated with deep brain stimulation, which has proven to be a relatively safe and effective treatment for the involuntary movements associated with Parkinson disease.

47. Question **1 points**

One night, a 75-year-old man was driving his wife home from a concert when he realized he was having great difficulty in discerning the lines designating the lanes. In fact, he almost hit another car when he accidently made a lane change where the road suddenly curved. His wife was greatly upset and insisted that he should not drive at night until he has been cleared by a doctor. Consequently, he made an appointment to see his primary care physician, who had been treating

him for type 2 diabetes and hypertension for over a decade; in providing a history, he informed the physician that he had become aware of gradually deteriorating vision; he stated that he had been noticing some difficulty in reading and that he also saw halos around light fixtures and headlights from oncoming cars during the night. Given this history, the most likely diagnosis would be which one of the following?

1. Retinal detachment
2. Age-related macular degeneration
3. Diabetic retinopathy
4. Hypertensive retinopathy
5. Cataract ✔

INCORRECT ✖

The correct answer is 5.

This patient has cataracts. Although cataracts can be congenital or acquired (due to diseases like diabetes mellitus or from medications such as long-term steroids), they most commonly occur with advancing age as “senile cataracts” (by the age of 80 years most person have had cataracts or at least precataract changes in their lens). Cataracts may be central or peripherally located (i.e., nuclear or cortical). Patients complain of a gradual loss of vision, blurred vision, and difficulty in driving a car at night, as in this case. Some of them will state that they have a problem with perceiving colors, while others may state that their vision has improved such that they require reading glasses with less power. This is often called “second sight,” and results from a nuclear cataract. On the other hand, patients who develop posterior subcapsular cataract will complain of deterioration in near vision. Although cataracts are often perceived as an advent normally associated with aging, they are due to denaturation of proteins in the lens; depending upon the major protein affected, they may be hard or soft, partial or more complete, unchanging or progressive. Additionally, any action or substance that promotes protein denaturation will accelerate cataract formation. Treatment is surgery, in which the old lens is removed and is replaced by a plastic one.

(Choice 1) presents as a sudden loss of vision, which is not the presentation in this case.

(Choice 2) is associated with progressive loss of central vision, which can appear gradually or suddenly.

(Choice 3) Although diabetes can promote diabetic retinopathy that may lead to deteriorating vision culminating in blindness, glare halos from headlights or light fixtures are not a feature.

(Choice 4) is associated with deteriorating vision, but once again, halos caused by glare from bright lights are not a feature.

48. Question **1 points**

A 45-year-old man who is a conforming Seventh Day Adventist who works as a technician in the laboratory of a Veterans Administration Hospital complains of increasing fatigue over the past few months. He has had to buy new trousers as his waist has been increasing in size, and he has difficulty in putting on his shoes. Recently, he also has noticed swelling of his breasts. These physical changes are most likely due to which one of the following causes?

1. Medication
2. Alcohol
3. Hepatitis A
4. Hepatitis B
5. Hepatitis C ✔

INCORRECT ✖

The correct answer is 5.

This patient has cirrhosis of the liver with portal hypertension. The expanding waist line is due to ascites, while swollen feet cause him to have difficulty in putting on his shoes. He has breast enlargement (gynecomastia) because the small amount of estrogen normally present in a male could not be metabolized by the failing liver. Other features would include palmar erythema and spider nevi. Approximately 20% of patients with chronic hepatitis C will end up with cirrhosis around 20 years after infection. Hepatitis C is most commonly transmitted via IV drug use. Other modes of transmission include sex, snorting cocaine, body piercing, tattoos, hemodialysis, blood transfusion, and upon occasion, an accidental finger stick by a laboratory worker. Unfortunately, there is no vaccine against hepatitis C, and the number of cases that would result in mortality and development of hepatocellular carcinoma is expected to increase threefold within the next one or two decades.

(Choice 1) medication is incorrect. Drugs such as isoniazid can induce hepatitis, but this is not very common, and nor is it common for medication-induced hepatitis to lead to cirrhosis.

(Choice 2) Alcoholic hepatitis is a major problem, but a conforming Seventh – day Adventist is not likely to be an alcoholic. Moreover, not all alcoholics with hepatitis develop cirrhosis of the liver. The incidence of chronic alcoholics who end up with cirrhosis of the liver is in the range of 10%–15%.

(Choice 3) is transmitted from the fecal–oral route and usually resolves spontaneously. Chronic hepatitis and cirrhosis does not occur, and immunization is available.

(Choice 4) can lead to cirrhosis of the liver, but the incidence is not as great as it is for hepatis C, and immunization against it is available.

49. Question **1 points**

On a hot August day in North Carolina, a 35-yearold Hispanic woman is seen by her primary care physician for progressive fatigue and a feeling of general malaise. She informs him that, over the past few days, she has been noticing difficulty in walking up a brush-covered slope leading to her home where she lives in the country, and she suspects she may be coming down with the flu. She has no history of chest pain, but did complain of some difficulty in breathing. She stated that she bruises easily. Apart from taking ibuprofen fairly regularly, which she purchases over the counter for arthritis, she has not been on any other any long-term medications. She had taken one ibuprofen a few hours prior to seeing her physician. She has no allergies. Physical examination reveals a moderately built and well-nourished female who has a low-grade fever and a yellowish tinge in the sclera. Her vital signs are within normal limits. She has normal heart sounds and no murmurs, gallops, or rubs. A few basal rales are present, but the lungs are clear otherwise. Examination of the abdomen is unremarkable except for mild tenderness in the right upper quadrant. She does have mild pitting edema in the pretibial area. The most likely cause for her problem is which one of the following?

1. Lyme disease
2. West Nile virus
3. Rocky mountain spotted fever ✔
4. Sarcoidosis
5. Q fever

INCORRECT ✖

The correct answer is 3.

Several factors provided in the case history are consistent with a diagnosis of Rocky Mountain spotted fever (RMSF), a disease caused by the transmission of Rickettsia rickettsii via a bite by a tick. These factors include the following:

■ The possibility of a tick bite is suggested by the statement that the patient had to walk up a brush covered slope in order to get home and that it was August, a time when ticks are most prevalent.

■ The patient lives in North Carolina (despite the name, only about 3% of all cases occur in a Rocky Mountain state, and most cases occur in the South Atlantic states with North Carolina having the greatest incidence).

■ Difficulty in breathing is likely an early sign of pneumonitis, as part of the acute respiratory distress syndrome (ARDS) often found in association with RMSF.

■ Pitting edema is present, which is a common sign of interstitial nephritis, a disease often induced by
R. rickettsii.

■ There is tenderness in the right upper quadrant, possibly an early sign of hepatomegaly, also a trait commonly associated with RMSF.

■ Yellowing of the sclera is likely an early sign of jaundice caused by excess hyperbilirubinemia due to hemolysis induced by RMSF.

As in the case described, initially, patients will have symptoms akin to influenza, such as fever, headache and myalgia. However, careful observation will discern early signs of some of the typical complications described above. After about the first week of the illness, a defining macular rash that progresses centripetally is usually seen. The initial rashes are at the wrists and ankles, and these may become petechial as they progress and as thrombocytopenia supervenes. In addition to the complications mentioned, a necrotizing vasculitis and uremia can also result. The diagnosis is confirmed by serology. Other laboratory finding suggestive of RMSF include elevated liver enzymes, thrombocytopenia, and hyponatremia. Treatment most typically is a tetracycline.

(Choice 1) Although Lyme disease is also transmitted by a tick, the incidence of new cases is highest in the New England states and is relatively low in North Carolina; even more significant to the contention that this is an incorrect choice is that early signs do not include hepatomegaly, respiratory problems, interstitial nephritis, or jaundice. Similarly, although interstitial nephritis and consequently pitting edema can be caused by several entities in addition to RMSF including prolonged use of nonsteroidal anti-inflammatory drugs (NSAIDs); some antibiotics, such as penicillin and cephalosporins; and infections other than RMSF, such as those caused by cytomegalovirus and streptococcus none of these factors is included among the choices nor do they cause the other symptoms described as associated with RMSF.

(Choice 2) West Nile virus is a flavivirus transmitted to humans by a mosquito bite and may cause encephalitis. The key features are neurological in nature, such as headache, fever, nuchal rigidity, stupor, and coma. Spastic paralysis and upper motor neuron signs and presence of lymphocytosis and elevated proteins on cerebrospinal fluid examination are present as well.

(Choice 4) is a disease of unknown etiology that primarily causes granulomatous inflammation in the lungs. It is more common in women than in men and among African American and Northern European populations. Being a systemic disease, it can target various organs, including the skin (causing erythema nodosum), heart, liver, spleen, kidneys, salivary glands, and peripheral nerves. Diagnosis is by histology, in which noncaseating granulomas are found on biopsy.

(Choice 5) is a rickettsial disease caused by Coxiella burnetii; it is transmitted by inhalation and not by vectors, as are many rickettsial diseases. It is usually seen in patients who are exposed to farm animals such as goats, sheep, and cattle. The patient presents with fever, headache, cough, and abdominal pain. Pneumonitis, hepatitis, and in some cases, endocarditis and encephalitis can ensue. The diagnosis is confirmed by serology.

50. Question **1 points**

A 20-year-old man comes to his family physician with a complaint concerning fainting attacks. He states that he has started going to the gym where he uses the treadmill and lifts some weights. He feels faint and sometimes actually passes out after being on the treadmill for some time; even more distressing, he almost always passes out soon after he begins to lift heavy weights. His father also had episodes of fainting; furthermore, he died following a heart attack at the age of 54 years. The

present patient has had a “double pulse” for a long time, and it has never bothered him. In fact, he likes to show off his special pulse to his friends. He denies chest pain, but does complain of some breathing difficulty. Upon examination, the physician observed a distinct cardiac murmur. This murmur would be expected to do which of the following?

1. Increase when he sits upright or when he squats
2. Decrease when he sits upright and remain unchanged when he squats
3. Increase when he holds his breath or when he squats
4. Decrease when he holds his breath or when he squats
5. Increase when he sits upright and decrease when he squats ✔

INCORRECT ✖

The correct answer is 5.

This patient has hypertrophic cardiomyopathy. Hypertrophic cardiomyopathy is a condition in which the left ventricular space is decreased due to the disproportionate hypertrophy of the interventricular septum. The restricted space reduces the amount of outflow. Performing a Valsalva maneuver or sitting upright increases the loudness of the murmur, whereas squatting reduces it. This is because a Valsalva maneuver or sitting upright decreases left ventricular filling and obstructs outflow. Likewise, sympathetic stimulation (as occurs during exercise) does the same, leading to symptoms such as syncope and even dangerous arrhythmias. Patients with hypertrophic cardiomyopathy have pulsus bisferiens, in which two peaks are felt, one following the other the “double pulse” described by the patient. Chest pain and dyspnea may be symptoms as well. Hypertrophic cardiomyopathy can be inherited as an autosomal dominant trait. It can result in dangerous arrhythmias and sudden death. His father must have died from sudden arrhythmias. The fact that the patient has the same disorder points to a genetic cause. A defibrillator should be implanted in patients who have a history of syncope, recurrent ventricular arrhythmias, or a family history of sudden death. **(Choices 1,2,3 & 4)** Since Choice 5 is correct therefore all other choices are incorrect.

TOP